

**A STUDY ON**  
**VAIGITHA VADHAM**

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## INTRODUCTION

Health is a ideal state of mind. When one is in health, his organ functions properly. And the life will be pleasant by the generation of optimistic thoughts & courage.

Even though health is a normal state of humans, it is not self-repetuating in order for its persistence health must be nurtured and cultivated. Certain conditions of living will be favourable to health. But deviation from this environment invites disease. So more emphasis must be laid on health rather on disease, and this must be intentional. But coin has two sides, and it will be less than realistic to state that thinking positively about health, will justify a person overlooking the dangers of disease.

Siddha system which is indigenous to this soil has been practised in Tamil Nadu. This system not only insists the cure of diseases, but it also paves way to attain perfection in various life arts such as philosophy, yoga, alchemy, medicine, wisdom and in particular the art of longevity.

The prince of mystics –Thirumoolar in his Thirumandhiram explained

"*ÁŪôÀÐ ¯¼øſſiö ÁÕó/¾É Ä;Îö*

*ÁŪôÀÐ ¯Çſſiö ÁÕó/¾Éî °;Öö*

*ÁŪôÀÐ þÉçſſiö Å;Ã; ¾çÖî,*

*ÁŪôÀÐ °; "Å ÁÕó/¾É Ä;ſÁ"*

Which correlates with the definition of health given by WHO. Their medical education was a mysterious process of initiation and apprenticeship in the top where they concentrated not only on the theory and doctrine, but also on the studying of diseases through observation.

In siddha, the theory of Panja Bootham has been applied to the origin of universe, and more specially to the development, physical function and pathological disorders of human body.

The derangement of three humours vadha, pittha, kapha results in disease. They described 4448 diseases on the basis of three thosha theory.

According to yugi vaithiya sindhamani “Vaighithavadham” comes under 80 types of vadha disease. The author selected Vaighithavadham (Bleeding disorder) for dissertation work. This is just a preliminary study. In the course of time with almighty grace further study will be carried out in the pathology of siddha system.

## **SIDDHA PHYSIOLOGY**

Nature is a material cause not merely of the outer universe but also of our body, with all its grosser and subtler divisions and components. The functioning of human body is governed by 96 specific faculties or tahthugas.

1. 7 Udal kattugal
2. 14 Vegangal
3. 6 Suvaigal
4. 3 Udal vanmai
5. 3 Udal thee
6. body constitution

Since siddha is based upon the saiva siddhantha philosophy, it also explains 96 thathugas

In Vedandha Thathuva Kattalai, the elements were described as,

“ $\bar{U}^{\frac{3}{4}}\hat{c}\hat{A}_i\bar{o}$   $\hat{a}^{\frac{3}{4}}_i\bar{c}$   $\$ \hat{A}_i$  “ $\tilde{A}\bar{o}^{\frac{3}{4}}_i\bar{o}$   
 $\bar{A}^{\div}$ ,  $\hat{c}\bar{y}\bar{E}$  /  $\hat{A}_i\bar{E}\hat{c}$  “ $\hat{A}\bar{o}\bar{D}$   $\bar{O}\bar{A}$  “ $\bar{E}\bar{o}^{\frac{3}{4}}_i\bar{o}$  . . .

The basic elements, Panchaboothas,

Mann – Earth, Neer – Water, Theyu – Fire, Vayu – Air, Aagayam – Ether.

As mentioned earlier, the bodily parts are made of these Panchaboothas. The parts were,

**Pori – 5**

Mei, Vaai, Kan, Mooku, Sevi

**Pulan – 5**

Saptham, Sparsam, Roopam, Rasam, Gantham

**Kanmaenthiriyam – 5**

Vaai, Kaal, Kai, Eruvaai, Karuvaai

**Gnanenthiriyam -5**

Vasanam, Kamanam, Dhaanam, Visarkam, Aanantham

**Anthakaranam – 4**

Manam, Buthi, Sitham, Agankaaram.

**Arivu – 1****Naadi -10**

Idakalai, Pinkalai, Suzhumunai, Siguvai, Purudan, Gaandhari, Athi,  
Alampudai, Sangini, Gugu.

**Vayu – 10**

Praanan, Abaanan, Viyaanan, Uthanan, Samaanan, Naagan, Koorman,  
Girugaran, Thaevathathan, Dhananjeyan

**Aasayam – 5**

Amarvaasayam, Pagirvaasayam, Salavaasayam, Malavaasayam,  
Sukilavaasayam.

**Kosam – 5**

Annamayakosam, Praanamayakosam, Manomayakosam,  
Vingnanamayakosam, Anandhamayakosam.

**Aaathaaram – 6**

Moolaathaaram, Swaathithaanam, Manipooragam, Anaagatham,  
Visuthi, Aakinai

**Malam – 3**

Aanavam, Kanmam, Maayai

**Mandalam – 3**

Gnayiru, Thingal, Agni

**Thodam – 3**

Vatham, Pitham, Kabam

**Eedanai – 3**

Porutpatru, Pudhalvarpatru, Ulagapatru

**Gunam – 3**

Sathuvam, Raasadham, Thaamasam

**Vinai – 2**

Nalvinai, Theevinai

**Raagam – 8**

Kaamam, Krotham, Ulopam, Moham, Madham, Marchariyam,  
Idumbai, Agankaaram.

**Avathai – 5**



Nanavu, Kanavu, Urakkam, Paerurakkam, Uyirpadakkam

### Udalkattugal

The main seven components built by our intake expand into human body. They are,

Saaram, Senneer, Oon, Enbu, Moolai, Kozhuppu, Sukkilam /Sronitham.

The energy from our food is distributed to this day by day and it takes seven days to enrich the 7 components.

### Vegangal

They were nothing but the reflexes, protecting mechanism of our body.

Vadham, Thummam, Siruneer, Malam, Kottaavi, Pasi, Neervetkai, Kasam, Ilaippu, Nithirai, Vaanthi, Kanner, Sukkilam, Swaasam.

### Suvaigal

"ÁñĪ¼Œ ĖĖ ¼ĒĪ, ĩø Ó"ĒĀĭ, Œ°÷Œ¼ĉŒ¼ĭø ĀŒŒĀĀĉĒĉŒ  
¼ĉñ½ĀĉĀŒ ĐĀ÷ŒĀĉĀŒ°Œ °¼ĭ, ¼ĉŒĀĭ¼ĭ÷ ¼ĒĀĉŒ ¼ĉ¼Āĭ Ó"ĒŒŒŒ  
±ñ½ĀĉĀ , °ŒŒñ¼ĭŒ ¼ñ½ĒĀĉø ,ĒĀĉ"½ŒĀĭø ±ŒĀĭ ŒĀ÷ŒŒŒ  
-ñ½ĀĉĀ «ŪĪ"ĀĀĉŒ ĀĉĒŒĀĉ/¼ŪŒ ĪŒ°ĉŒ¼÷ -"ĀŒ¼ Ā"ĒŒĀ"  
- ĀŒŒĐĀŒ ¼ĒĉŒĀĭ¼ø

The sense of taste explained here is six types and these can be sensed rightly only when the body is in the healthy condition.

They are, Inippu, Pulippu, Uvarppu, Kaarppu, Kaippu, Thuvarppu

These must be taken in a correct proportion for healthy living, since these have impact over the humours of the body.

### **Udal Vanmai**

It speaks about one's reason for having healthy contour of the body. They are, Iyarkai vanmai, Seyarkai vanmai and kaala vanmai.

### **Body Fires**

These are the main anabolic activators to supply energy to the demands of the body by the process of digestion and various activities.

Samanakini, Mandhaakini, Dheekshanaakini, Vishamaakini.

### **Body Constitutions**

During fertilization, the humour, determined by dietary and environmental factor determine the constitution and character of the individual.

**“நோய் நாடி நோய்முதல்நாடியது தணிக்கும்  
வாய்நாடி வாய்ப்பச் செயல்”**

Noi → means “Disease”

Naadal → means “to know the cause”

This kural implies that more importance has to be laid on to the disease, its cause and then only the treatment of disease.

**“மதித்திடற் கருமை வாய்ந்த  
மாண் பரிகார மெல்லாம்  
துதித்திட வுணர்ந்தானேறு  
துகளரப் பிணியின்றன்மை  
பதித்திட வுணரானாகிற்  
பயனுறா னாகலானே  
விதித்திடு பிணித் திறத்தை  
விளம்புது முதற்கண்முன்னே”**

**- சிகிச்சா ரத்ன தீபம்**

The 2<sup>nd</sup> poem indicates the importance of pathology than drug knowlege.

Siddha pathology explains the diseased condition of the body with respect to the changes in the humours, due to various factors, such as

1. Dietary changes

2. Environmental changes
3. Habit
4. Immoral changes
5. Seasonal Changes
6. Suppression of reflexes (veganga)
7. Hereditary factors.

These factors cause diseases in man and the symptoms are depicted according to the humour affected.

### **Diet**

According to siddha system, the daily food intake comes under 6 main tastes as இனிப்பு, புளிப்பு, உப்பு, துவர்ப்பு, கைப்பு, கார்ப்பு. Any alteration in the dietary habits affects the taste which finally affects the 3 humours and are depicted in the form of disease.

“ புளி துவர் விஞ்சங் கறியால் பூரிக்கும் வாதம்  
செறி யுவர் கைப்பேறில் பித்துச் சீறும் - கிளமொழியே  
கார்ப்பிணிப்பு விஞ்சில் கபம் விஞ்சம்”

i.e.,

Vadham → Increases with increase in sour and astringent tastes

Pitham → Increases with increase in salt and bitter tastes.

Kabam → Increases with increase in sweet and punget tastes.

### **Environment**

Environment also plays a vital role in determining disease. Ancient Tamilians classified the land into 5 types which are

1. Kurinji
2. Mullai
3. Marutham
4. Neithal
5. Pallai

### **Kurinji**

Person dwelling in this land will be affected with fever, anemia, liver enlargement, kaba diseases etc.,

*“குறிஞ்சி வரு நிலத்திற் கொற்றமுண்டி ரத்தம்  
உறிஞ்சி வரு சுரமு முண்டாம் - அறிஞரைக்  
கையமே தங்குதரத் தானம்வல்லை யுங்கதிக்கும்  
ஐயமே தங்கு மறி”*

Similarly,

Mullai → Vadha & Pitha diseases are predominant

Marutham → Favourable place for dwelling

Neithal → This land is more prone to vadha disease, elephantiasis and hepatomegaly.

Palai → Abode of all ailments

It is inferred that the environment alone with its humidity, fertility, nature of soil, its capacity, population, plays a vital role in man's internal environment, immunity, the defence and also for the organisms causing diseases in endemic and epidemic situations.

### **Habits**

They are the deeds involving one's mental and physical activity. Good deeds and their consequences keep the individual fresh and healthy whereas, the notorious deeds lead them to continued stress, reduced immunity and make embodiment of diseases.

### **Immoral Activities**

In this modern world, there is a wide opportunity for the human to derail from the disciplined path. This paves way for the dreadful diseases.

### **Season**

Seasonal variations are depicted in a major & 6 minor forms. The relation between them and the humours are

Season	Period	Mukkutram		
		V	P	K
Kaar	Avani & Purataasi	*	☼	
Kudhir	Aippasi & Kaarthigai	●	*	
Munpani	Maarghzzhi & Thai		●	
Pinpani	Maasi & Panguni			☼
Ilavenil	Chittirai & Vaigasi			*
Mudhuvenil	Aani & Aadi	☼		●

- Normal State

✿ Alternation from normal state (தன்னிலை வளர்ச்சி)

\* Aggrevation and spread to the other humour (பிறநிலை வளர்ச்சி)

### **Suppression of reflexes**

Physiological urges when inhibited causes disease.

The 14 physiological urges are

### **Vadham (Abana Vayu)**

Chest pain, peptic ulcer, abdominal pain, body ache, constipation, oliguria and indigestion.

### **Thummal**

Headache, facial pain, back pain, pain over the sense organs etc.,

### **Siruneer**

Ulcers in the urethral orifice, joint pain, urinary tract infections etc.,

### **Malam**

Calf muscle pain, headache, general debility, flatulence and other diseases.

### **Kottavi**

Indigestion, contractures in the face.

### **Pasi, Neervetkai**

Constitution of the body is totally disturbed, emaciation.

### **Kasam**

Chest disorders supervene.

## **Ilaippu**

Ulcer and other major diseases.

## **Nithirai**

Heaviness of the head, eyes, watering of the pain, deafness, speech disturbances.

## **Vaanthi**

Urticaria, skin diseases, toxic manifestations, anaemia, eye disease.

## **Kaneer**

Eye diseases, headache, sinusitis and heart diseases.

## **Sukkilam**

Joint pain, fever, chest pain, difficulty in micturition.

## **Swaasam**

Cough, abdominal discomfort, anorexia.

By these factors, the normal picture of the humours are shuffled and are expressed as diseases.

## **Hereditary factor**

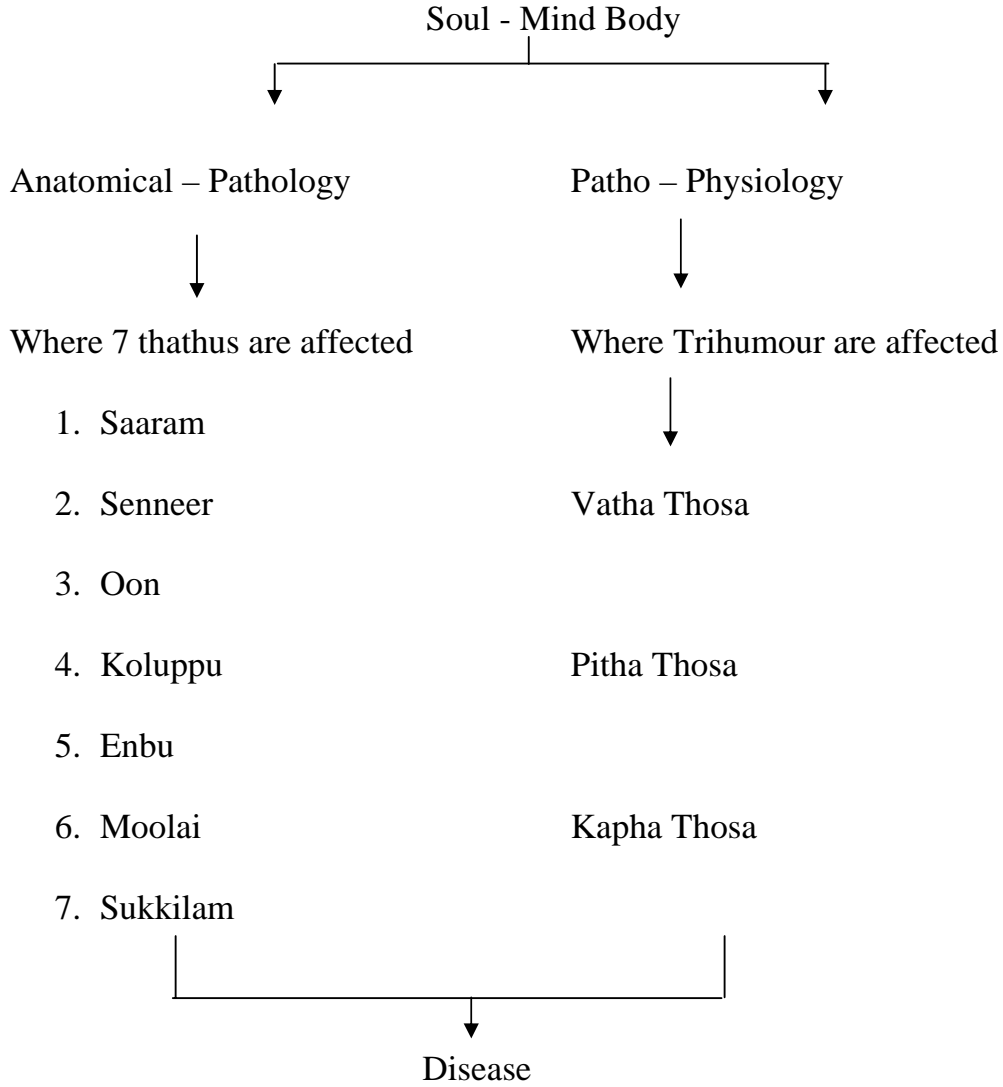
Heredity also plays an important role in the cause of disease. This is depicted by our

**“பேறுயிளமையின்பம் பிணி மூப்பு சாக்காடு**

**ஆறுங்கருவிலமைப்பு”**

All these are reflected in the form of disease, which affects mind and body.





### Envagai Thervugal (8 tools of diagnosis)

The above said features can be located by the unique method of envagai thervu

1. Examination of Pulse (நாடி)
2. Examination of touch (ஸ்பரிசம்)
3. Examination of tongue (நாக்கு)
4. Examination of complexion (நிறம்)
5. Examination of voice (மொழி)

6. Examination of eyes (விழி)
7. Examination of faeces (மலம்)
8. Examination of urine (மூத்திரம்)

These eight entities can be executed by the methods of poriyaal aridhal, pulanal aridhal, vinavudhal, and these are explained under

**“அளவை காண்டல் கருதல் உரை அபாவம் பொருள் ஒப்பாறென்பர்  
அளவை மேலும் ஒழிப்புண்மை யைதிகத் தோடியல்பென் நான்  
களவை காண்பர் அவையிற்றின் மேலும் அறைவர் அவையெல்லாம்  
அளவை காண்டல் கருதல் உரை என்றும் முன்றிலடங்கும்”**

**- சிவசித் அளவை**

By interrogating feeling, seeing the symptoms and signs are heard and examined. After examining, it must be compared, excluded and at last the final diagnosis is to be arrived.

### **Mei**

It deals all about the changes in the skin (ie) tactile sensation, the warmth, the chillness, sweat, numbness, fissures, plaques, papules, ulcers, inflammation etc.

### **Niram**

The normal colour of each humoral body is explained. If there is any change from normal (ie) is colour of eyes, tongue, mucous membrane, any erythema, hypo (or) hyperpigmentaion in the skin, they are dealt under this.

**Thoni**

This not only explains the tone of speech but also the changes in modulations, pitch, sound, fluency, stammering, difficulty in articulating, repetition, listening, answering speech, associated with breathing difficulties etc.

**Vizhi**

The view on one's eyes stretches all sides. It deals about the vision changes such as loss of vision, blurred vision, changes in visual perception, movements eye lids & eye balls, colour of conjunctiva and growth lacrimation, dryness, contractions, congenital defects are also specified under this examination.

**Naa**

It reflects the disease and so it gains importance in examining. The tongue is seen for the colour, shape, size, coating, fissures, growth, surfaces, sensations of taste and also salivary secretion.

**Malam**

The metabolic end products of our food after completing its work of supplying energy is expelled from the body as faeces. And thus any change in the colour, consistency, frequency, amount, components of motion exhibit the disease.

## Moothiram

Urine plays an important role in revealing the diseased state in the form of changes in colour, specific gravity, odour, frequency, froth and deposits.

**“வந்த நீர்க்குறி எடை மணம் நுரை எஞ்சலென்**

**ரைந்திய லுளவ வையறை சூது முறையே”**

## Neikuri

**”அருந்துமா றிரதமும் அவிரோ தமதாய்**

**அ.:கல் அலர்தல் அகாலவூண் தவிர்ந்தழற்**

**குற்றள வருந்தி உறங்கி வைகறை**

**ஆடிக் கலசத் தாவி-யே காது பெய்**

**தொருமுகூர்த் தக்கலைக் குட்ப டுநீரின்**

**நிறக்குறி நெய்க்குறி நிருமித்தல் கடனே ”**

This is an unique and special methodology in determining the diseases. The early morning first voided mid stream urine is taken in a glass bowl. A drop of gingelly oil is let into its surface. It forms many shapes due to the surface tension exhibited by the urine. If there is any change in the body metabolism, there will be alterations in the components of urine and thus the surface tension depicting various structures.

Vadha diseases – Rays of snake (பாம்பை போல் நீண்டல்)

Pitha diseases – As a ring (மோதிரம் போல்)

Kaba diseases – Stands as a pearl (பரவாமல் முத்துபோல்)

## Naadi

It is the diagnostic entity and felt in the radial artery with the three fingers, fore finger (vadham), middle finger (pitham) and ring finger tips (kabam). Ratio is 1: ½ : ¼. For male it is felt in right hand and for female, left. The alterations denotes and confirms the diagnosis made by other 7 examinations.

*“பேர்ந்திடவே சகலருக்கும் கரத்தினாடி*

*பேசினார் பிரமமுனி பேசினாரே”*

*“முப்பிணி மருவி முனிவுகொள் குறிப்பை*

*தப்பாதறியுந்த தன்மையும் வாத*

*பித்தவையப் பிரிவையுமவைதாம்*

*ஏறியிறங்கி இணைந்துக் கலந்து*

*மாறி மாறிவருஞ் செயற்கையாற் பிணி*

*நேர்மை யறிந்து நீட்டுமருந்தே*

*சீரியதா மெனச் செப்புவர் சித்தரே”*

Which indicates the importance of Naadi.

## Udal Thathus

When the food materials are digested, they are absorbed as saaram

## 7 Thathus

When food enters 1<sup>st</sup> day

Rasam	-	Plasma		
2 <sup>nd</sup>	-	Ratham	-	Blood
3 <sup>rd</sup>	-	Mamisam	-	Muscular tissues
4 <sup>th</sup>	-	Koluppu	-	Fat and Lymph
5 <sup>th</sup>	-	Enbu	-	Bones and cartilages
6 <sup>th</sup>	-	Moolai	-	Bone marrow
7 <sup>th</sup>	-	Sukkilam	-	Reproductive fluids

If there is any alteration in the process, it is reflected as disease which is due to the derangement of 3 humours.

### **1. Saaram (Rasam)**

Rasam is the essence of digested food which circulates all over the body by “Vijana Vaja”. The main functioning of Rasam is nourishing and production of food.

#### **Increased state**

It indicates provoked kapha symptoms like

1. Excessive salivation
2. Anorexia
3. Body ache, cough, excessive sleep etc.,

#### **Decreased state**

- Wasting of muscles
- Sound intolerance
- Generalized weakness etc

## **2. Senner**

Senner is produced from Rasa thathu. It is responsible for substances of life and provides colour and complexion to the skin.

### **Increased State**

- Haemangiomas
- Spleenomegaly
- Leprosy
- Jaundice
- Mental disorders
- Blood dyscariasis
- Hyperpigmentation

### **Decreased state**

- Anaemia
- Dry skin
- Nervous weakness
- Desire for sour foods

## **3. Oon (Mamisam)**

They are muscular tissue in the body. It is produced from Senner.

### **Increased state**

- Tumours
- Carcinoma

- Goitre
- Cyst
- Musculature

#### **Decreased state**

- Wasting
- Dryness
- Cracking sound in joints etc

#### **4. Koluppu (Medas)**

It means, circulating fat (lymph) and deposited fat (Adipose tissue)

#### **Increased state**

- Associated with mamsa disorders, like tumours
- Obesity
- Increased musculature with deposition of lymph (or) fatty tissue.
- Hypercholesteremia

#### **Decreased state**

- Wasting of muscles
- Decreases stability of hip joint and other joints
- Lethargy (or) sluggishness.

#### **5. Enbu (Asti)**

It indicates bones and cartilages and gives the structural frame work to the body.



**Increased state**

- Hypercalcinosis
- Extra teeth formation
- Hypertrophy of bone

**Decreased state**

- Osteoporosis
- Rickets

**6. Moolai**

It denotes bone marrow, nerves, grey and white matter of central nervous system. (Nervous tissue).

**Increased state**

- Bone and joint diseases
- Ulcers
- Feeling of weariness in eyes and body.

**Decreased state**

- Demyelination
- Osteomyelitis
- Delusion
- Giddiness

**7. Sukila Thathu (Sukkelum)**

It denotes reproductive system of both male and female.

### Increased state

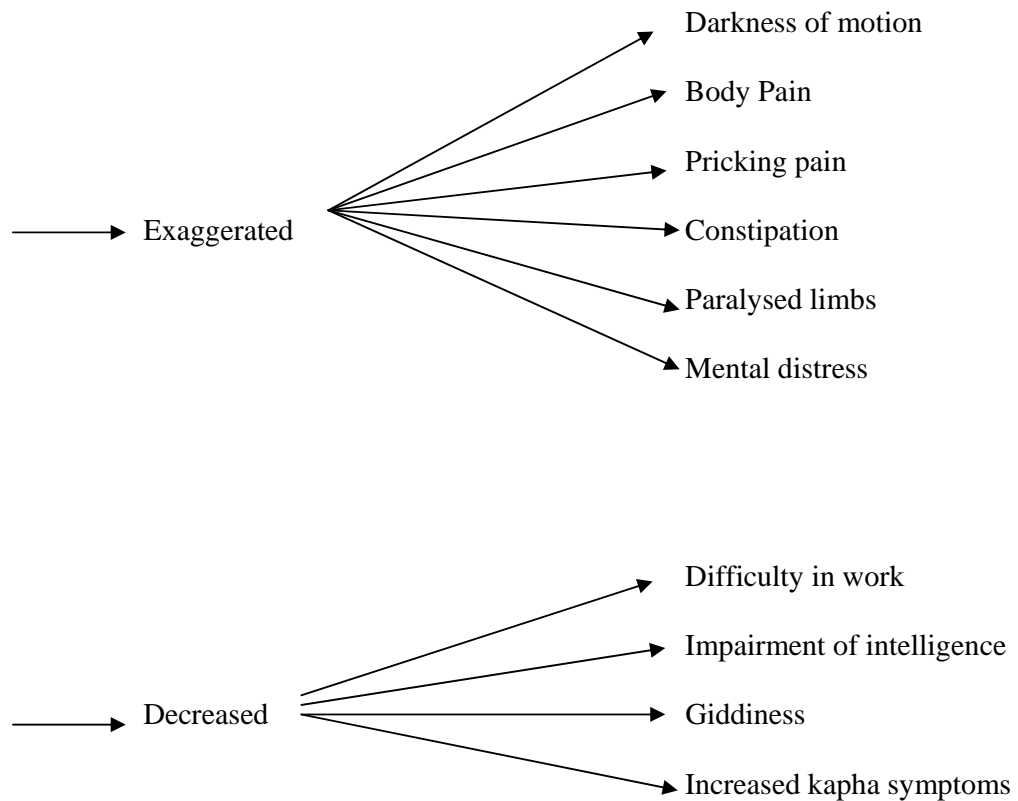
- Stone in urethra
- Increased sexual desire

### Decreased state

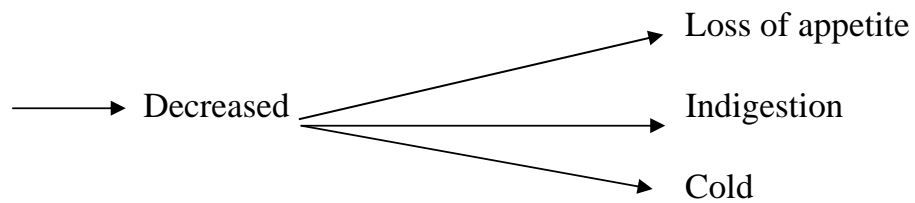
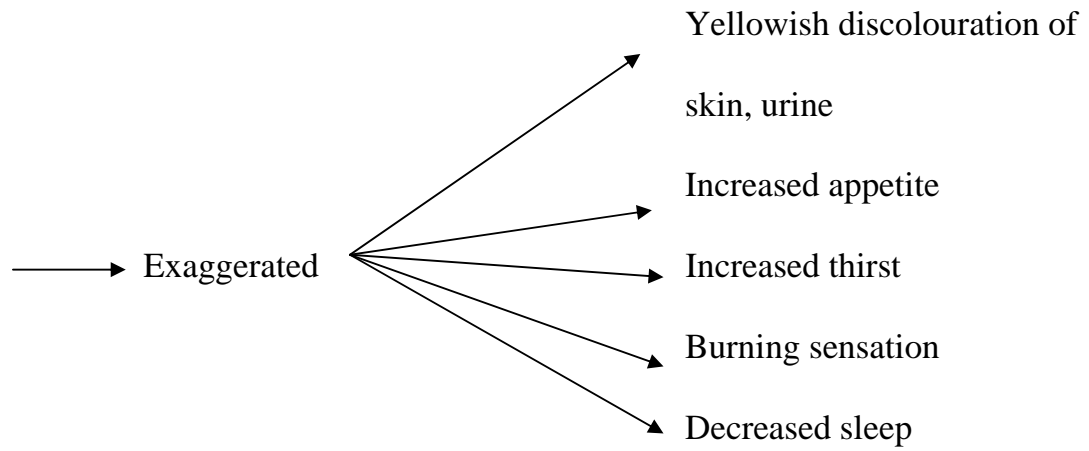
- Impotency
- Infertility
- Weakness

### De-Arrangement of 3 humour

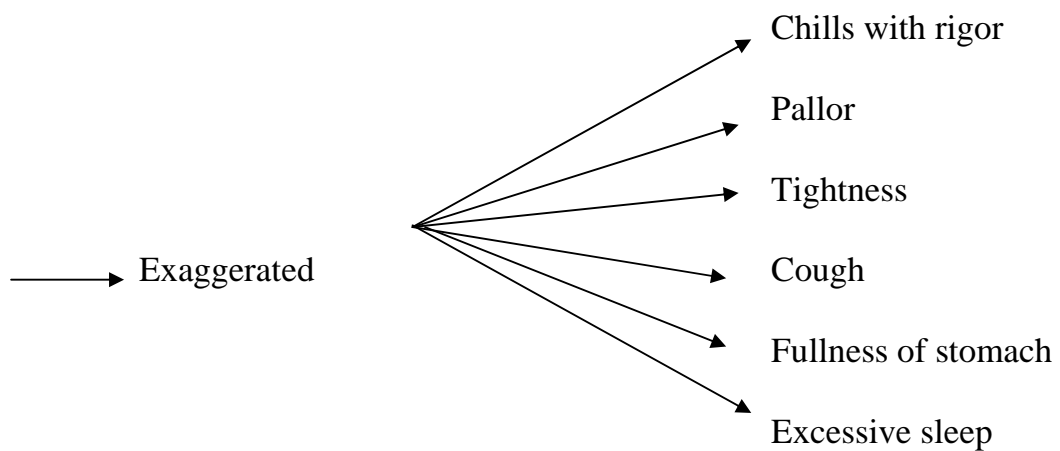
#### 1. Vatha – Thodam:

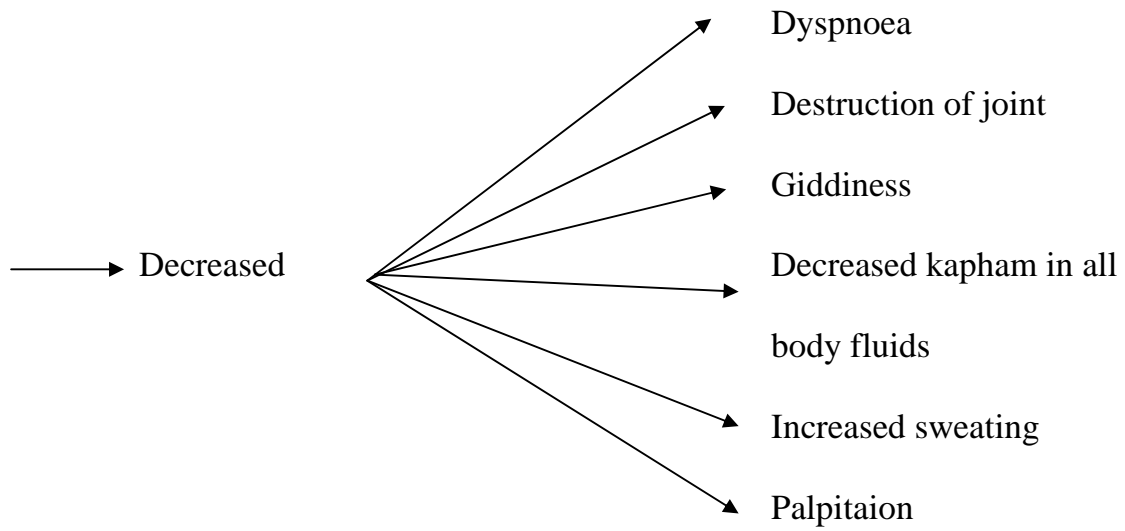


## 2. Pitha thodam



## 3. Kapha thodam





This is just a introductory work of the author, trying to evaluate the pathology of the symptoms depicted in “Vaigidha Vatham” (One of the vadha disease) in Yogi Vaidhya Chindhamani.

## AIM AND OBJECTIVE

**“பொற் றாமரையான் புனைமெய் யரண்காக்கும்**

**பொற்றா மரையான் புகல்வாதன் - பொற்றாம்**

**வளவினிலே யாக்குரம்பை மண்ணென்ன மன்ன**

**வனவினிலே யாக்கும் வலி”**

**- தேரையர் யமகவெண்பா**

சரீரத்தில் உயிர் தங்கி நிலைபெறச் செய்யும் வாதமானது இச்சரீரமாகிய கோட்டையை அரசாட்சி புரியும் வேந்தன் என்று வாதத்தின் பெருமையை பற்றி தேரையர் கூறியுள்ளதாலும்

**வாதமே கதித்த போது வந்திடும் வியாதி மேலும்**

**வாதமே கதித்த போது வல்லுடல் மெலிந்து கொள்ளும்**

**- அகத்தியர் சிகிச்சாரத்னதீபம்**

The derangement of vadha humour aggravated the diseases, and produce hazardous effects.

The author has selected Vaigithavadham which comes under vadha diseases. Vaigithavadham, the most common inherited human bleeding disorder shows a high incidence in the population. The main aim of this

study is to evaluate the pathology of the disease “Vaigithavadham” with the help of siddha and modern parameters.

The study includes

- to collect reviews in siddha literature
- efficacy of siddhars diagnostic tools
- to have an idea about the prevalence of the disease with respect to age, sex and family
- to know how the disease alters normal condition under mukkutram, pori, pulangal, udal kattugal, envagai thervugal, neerkuri, neikuri.
- to correlate etiology, symptoms, diagnosis with modern system of medicine.
- to use the modern diagnostic parameters to confirm the disease.

## READING LINES BETWEEN YUGI'S POEM

**Reading lines between yugi's poem:-**

**“ஆமென்ற வீங்கினதோர் விடத்தில்ரத்தம்”**

Haematoma formation at a particular place due to clotting factor deficiency.

**“அழுத்தமாய் திரண்டுமே எங்கும் பாய்ந்து**

Firm in character and rounded in appearance which means the inflammatory reaction at a particular place.

**“ஓமென்று ஒட்டியே திரண்டிருக்கும்”**

The rounded mass appears to be shining in its appearance

**“உறுதியாய்த் தொட்டுடனே மெத்தென் றாகும்”**

Pain on touch which is due to the mediators of inflammation.

**தேமென்ற தேகமெங் கணுமு சுக்கும்**

The changes may occur at any part of the body.

**சீறியதோ ரிருமலோடு காய்ச்சலுண்டாகும்**

Presence of cough and fever, cough, is due to the reduced hum oural or cellular response to infections in a generally debilitated patient. Secondly, it may occur due to respiratory tract haematoma Diaphragmatic haemotoma.

Fever produced by cytokines is due to the release of prostaglandins in the hypothalamus.

**பாமென்ற படந்தனிலே திமிருண்டாகும்**

Formed haematoma, compresses the adjacent nerves which leads to symptom like numbness of the body.

### **பாரமாய் வைகிதமாம் வாதந்தானே**

All these symptoms indicate the disease Vaigitha vadham.

The stanza analyses the signs and symptoms of the disease, starting from haematoma, its characteristic features like swelling which is firm in nature, pain and fullness. It clearly depicts that the disease can occur in any part of the body. The stanza also highlights other clinical features like fever, cough & numbness of the body.



## DETAILED PATHOLOGICAL VIEW OF THE

### DISSERTATION TOPIC

Pathological View of dissertation topic:

ஆமென்ற விங்கினதேர் விடத்தில் ரத்தம்  
அழுத்தமாய்த் திரண்டுமே எங்கும் பாய்ந்து  
ஓமென்று ஓட்டியே திரண் டிருக்கும்  
உறுதியாய்த் தொட்டுடனே மெத்தென் றாகும்  
தேமென்ற தேகமெங்கனுமு சுக்கும்  
சீறியதோ ரிருமலோடு காய்ச்சலுண்டாகும்  
பாமென்ற படந்தனிலே திமிருண்டாகும்  
பாரமாய் வைகிதமாம் வாதந்தானே”

This Stanza highlights clinical features of the disease, as follows:

1. Bleeding with the formation of haematoma.
2. Characters of haematoma.
3. Cough
4. Fever.
5. Complications of disease like ischaemic damage to nerves.

*ஆமென்ற வீங்கினதோர் விடத்தில் ரத்தம்*

*அழுத்தமாய்த் திரண்டுமே எங்கும் பாய்ந்து*

*ஓமென்று ஓட்டியே திரண் டிருக்கும்*

*உறுதியாய்த் தொட்டுடனே மெத்தென் றாகும்*

### **Bleeding with formation of haematoma:**

Generally the blood flows inside the closed circuits of the blood vessels which are arteries, arterioles, capillaries, veins and venules. Physiologically, blood is in constant motion inside the circuits. The escape of blood from the blood vessel is called haemorrhage and this extravasation of blood in to the tissues results in swelling known as haematoma.

### **Excessive bleeding may result from:**

1. Increased fragility of vessels.
2. Platelet deficiency or dysfunction.
3. Derangements in coagulation.
4. Combination of above three.

### **Increased fragility of Vessels:**

Disorders with in this categories usually do not cause serious bleeding problem: Most often they induce small haemorrhages, petechiae and purpura in the skin or mucousmembrane.

**Platelet deficiency:**

Reduction in platelet number constitutes a characterized bleeding most often from small vessels, where the bleeding is usually associated with small petechiae.

**Derangement in coagulation:**

The deficiency of one of the knowing clotting factors results in bleeding disorder. The bleeding in this condition differs from the other two with the uncommon appearance of petechiae or purpura. More often bleeding manifest as the development of large haematomas followed by injuries.

So, the author concludes the disease is due to deficiency of clotting factors.

The activated factor IX, acting in concern with activated factor VIII and with platelet, phospholipids and platelet factor III from traumatized platelets, activates factor X. So it is clear that when factor VIII, IX is deficient this step is defecit. And further process won't occur.

In person with deficient factor VIII &IX the process of

1.Vaso constriction.,

2. Platelet plug formation

occurs, but they lack in the process of conversion of fibrinogen to fibrin, resulting in bleeding which in turn leads to the formation of haematoma.

Extravasations of blood in to the tissues with resultant swelling known as haematoma. As soon as the haematoma is formed the inflammatory

response sets in, which is due to changes in the vascular flow and calibre of small blood vessels in the injured tissue. The sequence of these changes are as follows,

- The immediate vascular response is of “Transient Vasoconstriction” of arterioles.
- Next follows “Persistent Progressive Vasodilatation” which involve mainly the arterioles and to less extent venules and capillaries. Vasodilatation results in increased blood volume in microvascular bed of the area which is responsible for redness and warmth.
- Progressive Vasodilatation, in turn, elevates the local hydrostatic pressure resulting in transudation of fluid in to the extra cellular space, thus forming the swelling.
- Stasis of microcirculation increases the permeability of micro vasculature which results in increased concentration of red cells, followed by Leucocytes Margi nation, where the Leucocytes move through the gaps between the endothelial cells into the extravascular space. All the above said features results in redness, heat and swelling.
- The numerous endogenous mediators of increased vascular permeability that part take in other process of inflammation like pain.

The inflammatory pain sets in and persists until the injury heals. Characters tickly stimuli in the injured area produce an exaggerated response “Hyperalgesia” and innoculo us stimuli such as touch, cause pain

“Allodymia”. Inflammation of any type causes the release of many different cytokines and several vasoactive amines like Histamine, Serotonin in the inflamed area. Many of these facilitate perception and transmission in cutaneous as well as dorsal horn. This is what causes the hyperalgesic and allodymia

### **தேமென்ற தேகமெங்கனுமு சுக்கும்**

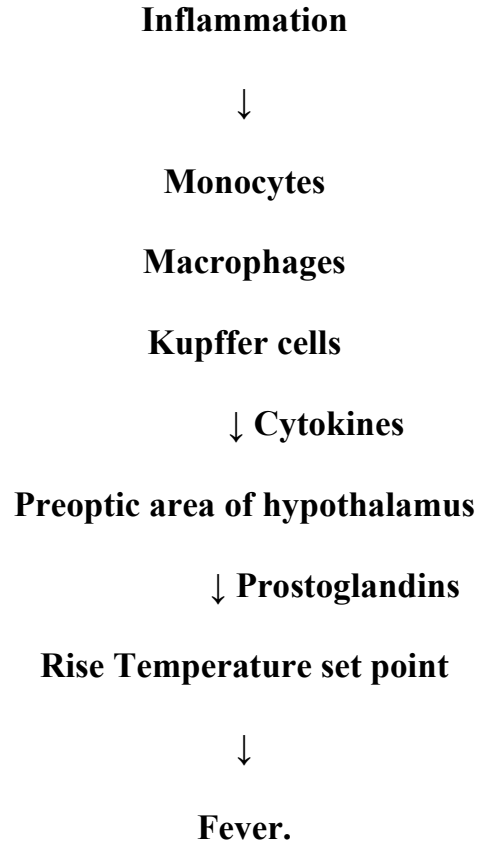
As the blood flows inside the closed circuit of the blood vessels, even a minor injury results in bleeding which occurs in any part of the body.

### **சீறியதோ ரிருமலோடு காய்பச்சலுண்டாகும்**

The reduction in blood volume due to bleeding leads to anaemia, with the following symptoms like lassitude, fatigue etc. Due to general debility the patients will have reduced cellular or humoral responses to infections. Hence infections of the respiratory tract will produce cardinal symptom like cough.

The bronchi and trachea are so sensitive, even upper respiratory tract bleeding, diaphragmatic haematoma, causes irritation, that initiates the cough reflex. The afferent impulses pass from the respiratory passages through the vagus nerve to the medulla. There on automatic sequence of events is triggered by the neuronal circuits of the medulla causing the cough reflex, so that the rapidly moving air usually carries with it the matter (i.e.) (haematoma) that is present in respiratory passage.

## **Pathogenesis of fever :**



Mediators of inflammation, cytokines (IL-II, IL-6,  $\beta$ -1FN,  $\gamma$ -IFN, TNF- $\infty$ ) act as endogenous pyrogens. These are polypeptides and the circulating cytokines act on OVLT (Organo Vasculosam of Lamina Terminalis) one of the circum ventricular organs. This in turn activates the preoptic area of hypothalamus. The fever produced by cytokines is due to the release of prostaglandin in the hypothalamus. PGE2 is one of the main prostaglandin that causes fever.

***பாமென்ற படந்தனிலே திமிருண்டாகும்***

***பாரமாய் வைகிதமாய் வாதந்தானே***

Muscle Haematoma when formed may extend to press on the nearby nerves with consequent numbness in that area. If the bleed does not resolve completely, recurrence may occur, leading to progressive muscle damage and ischemic damage to the nerves. For example, a large psoas bleed may extend to press on the femoral nerve with Para in the thigh and weakness of Quadriceps and the patient is left with weakness in the leg.

## **EVALUATION OF DISSERTATION TOPIC**

### **MATERIALS AND METHODS**

The clinical study on vaigitha vadham was carried out at the post graduate department of noi naadal branch in govt siddha medical college, palayamkottai.

### **CASE SELECTION AND SUPERVISION**

The following 12 cases selection and supervision was done in Peace Health Centre, Tirunelveli according to the clinical features of vaigithavadham(Haemophilia), as mentioned by yugi vaithiya chinthamani.

The patients are carefully examined systematically under the supervision of the professor and lecturer of the post graduate noi naadal department.

The detail history of the present and past illness and family history were observed. Typical picture of 12 cases was evaluated under siddha and modern parameters.

### **EVALUATION OF CLINICAL PARAMETERS**

The detail history and clinical features of the patients were taken carefully.

The clinical history contains,

- Detail history of present and past illness



- Personnel and family history

Clinical features of vaigitha vatham are

- Bleeding with formation of haematoma
- Swelling with pain
- Cough
- Fever
- Numbness

Study of siddha clinical diagnosis

Modes of investigation, the cases are

1. Poriyal arithal
2. Pulanal arithal
3. Vinathal
4. Mukkutra nilaigal
5. Udal kattukal
6. Envagai thervugal

### **The clinical investigation**

For further detailed study about the disease the following laboratory investigations was done in all cases.

1. Haematology
2. Bleeding time
3. Clotting time
4. Prothrombin time

5. Partial thromboplastin time
6. Factor assay
7. Total count
8. Differential count
9. Haemoglobin
10. Erythrocyte sedimentation rate

### **Bio chemical**

Blood sugar

Blood urea

### **Urine**

Albumin

Sugar

Deposits

### **Motion**

Ova

Cyst

### **Other test**

Xray

Ultra sonogram

## OBSERVATION AND RESULTS

Results are observed with respect to the following aspects.

1. Age and Sex reference
2. Family history
3. Mukkutraniilai
4. Udal thathukkal
5. Envagai thervugal
6. Clinical features
7. Laboratory findings

### Age and Sex reference

Age	Sex		No of cases
	Male	Female	
Upto 10yrs	3	-	3
10 – 20 yrs	3	-	3
20 – 50 yrs	6	-	6

## Family History

Case Number	Family history
1	Positive
2	Negative
3	Positive
4	Negative
5	Positive
6	Positive
7	Negative
8	Positive
9	Positive
10	Positive
11	Negative
12	Positive

## Mukkuṭranilai

### a. Derangement of vadham

S. No	Types of vadham	No of cases affected	Changes
1	Pranan	-	-
2	Abanan	-	-
3	Viyanan	12	Difficulty in movements of affected areas
4	Uthanan	3	Cough
5	Samanan	12	It's balancing function is disturbed
6	Nagan	-	-
7	Koorman	-	-
8	Kirukaran	-	-
9	Devathathan	-	-
10	Dananjeyan	-	-

### b. Derangement of pitham

S. No	Types of Pitham	No of cases affected	Changes
1	Anar Pitham	-	-
2	Ranjaga Pitham	5	Paleness of the tongue
3	Sadhaga Pitham	12	Difficulty in doing routine works
4	Aalosaga Pitham	-	-
5	Prasagapitham	12	Colour changes in the skin at the site of injury

### c. Derangement of Kabham

S. No	Types of Kabham	No of cases affected	Changes
1	Avalam pagam	7	Its balancing function is disturbed
2	Kilethagam	-	-
3	Bothagam	-	-
4	Tharpagam	-	-
5	Santhigam	7	Difficulty in movements

### 3. Udal Thathukkal

S. No	Udal Thathukkal	No of cases	Changes
1	Saaram	12	Malaise, dizziness
2	Senneer	12	Stagnation of blood in the site of injury
3	Oon	-	-
4	Kozhuppu	-	-
5	Enbu	-	-
6	Moolai	-	-
7	Sronitham	-	-

## The picture of Envagai Thervugal

<b>Case No</b>	<b>Sparisum</b>	<b>Naa</b>	<b>Niram</b>	<b>Mozhi</b>	<b>Vizhi</b>	<b>Malam</b>	<b>Moothiram</b>	<b>Naadi</b>
1	A	N	A	NA	N	N	A	VK
2	A	N	A	NA	N	A	A	PV
3	A	N	A	NA	N	N	N	VP
4	A	A	A	NA	N	N	N	VP
5	A	A	A	NA	N	N	N	VP
6	A	A	A	NA	N	N	N	VK
7	A	A	A	NA	N	A	N	VK
8	A	N	A	NA	N	A	N	VP
9	A	N	A	NA	N	N	N	VP
10	A	N	A	NA	N	N	N	VP
11	A	A	A	NA	N	N	N	VP
12	A	N	A	NA	N	N	N	VK

N – Normal, NA – Not Affected, A – Affected, VK – Vatha kabham

VP – Vadha Pitham, PV - Pithavadham

### Clinical Feature

S. No	Clinical Features	No. of cases
1	Bleeding with formation of haematoma	12
2	Characters of haematoma	12
3	Fever	4
4	Cough	3
5	Numbness	6

### Factor assay – Report

Case No	Age	Factor – Report
1	23	VIII - < 1%
2	20	VIII - 40%
3	10	F.VIII - 10 %
4	9	IX - <1%
5	3	VIII - 56%
6	22	VIII - 4%
7	24	VIII - 0.5%
8	45	VIII - 1.47%
9	19	VIII - <1%
10	19	VIII - 20%
11	8	VIII - 4%
12	12	VIII - 3%



## Laboratory Investigation

Case No	BT	CT	PT	PTT	PT count
1	3 min	7 min	12.5sec	32 sec	1.2
2	2 min	4 min	12 sec	30 sec	1.6
3	2 min	4 min 45 sec	15 sec	28 sec	1.4
4	1'45"min	6.00 min	14 sec	30 sec	1.2
5	12'30"min	7.30 min	12 sec	33 sec	1.2
6	2.00 min	8.00 min	15 sec	34 sec	1.5
7	6 min 10sec	12 min 55sec	12.3 sec	37 sec	1.8
8	2 min	45min	11.3 sec	64.9 sec	1.6
9	1'40 min	6.20 min	12 sec	36 sec	1.8
10	2 min	6 min	11.3 sec	33.5 sec	3.57
11	2 min	8.00 min	12 sec	30.5 sec	1.2
12	1' 40 min	7 min	13 sec	38 sec	11

### Normal values

Bleeding time – 2. – 4 min

Clotting time – 3 – 6 min

Prothrombin time – 0 – 27 sec

Partial thromoplastin time – 0 – 25 sec

Case No	Blood				ESR		Hb	Bio - Chemical		Alb	Sug	Dep	Ova	Cyst
	TC cells cumm	DC Cells			1/2 hr mm	1 hr mm		Sugar msg%	Urea msg%					
		P%	L%	E%										
1	9000	50	40	10	8	16	12	120	25	-	-	-	-	-
2	10000	52	38	10	8	18	13	121	25	-	-	-	-	-
3	19600	91	6	3	2	6	11	187	25	-	-	-	-	-
4	7000	46	52	2	8	20	9.8	110	23	-	-	-	-	-
5	15400	26	68	6	4	12	9	-	-	-	-	-	-	-
6	10000	60	36	4	16	30	10.5	120	25	-	-	-	-	-
7	9000	56	41	3	30	58	10	70	27	-	-	-	-	-
8	7300	69	28	3	16	30	12	110	25	-	-	-	-	-
9	8200	56	40	4	20	40	12.4	90	23	-	-	-	-	-
10	1300	58	34	8	28	50	12	86	24	-	-	-	-	-
11	11000	65	31	4	4	10	10	80	20	-	-	-	-	-
12	10000	60	37	3	12	20	11	80	23	-	-	-	-	-

# **THEORETICAL VIEW OF DISSERTATION TOPIC IN**

## **MODERN ASPECT**

### **Haematology**

#### **Blood**

Blood is a vital connective tissue pumped by the heart through all the arteries, veins and capillaries.

#### **Properties of blood**

##### **Colour**

Blood is an opaque fluid and it is red in colour. Arterial blood is scarlet red and venous blood is purple.

##### **Volume**

The volume of blood in a normal adult is 5 liters.

##### **Reaction and PH**

Blood is slightly alkaline and its PH in normal condition is 7.4

##### **Specific gravity**

The specific gravity of total blood : 1.052 to 1.06

The specific gravity of blood cells : 1.092 to 0.101

The specific gravity of plasma : 1.022 to 1.026

##### **Viscosity**

Blood is five times more viscous than water. It is mainly due to red blood cells and plasma proteins.

## **Composition of blood**

Blood contains liquid portion known as plasma and the formed elements called the blood cells.

1. Plasma
2. Blood cells

### **Plasma**

The plasma of the blood is formed by 91 to 92 % of water and 8 to 9 % of solids are the organic and the inorganic substances.

### **Organic substances of the plasma**

The following are the organic substance of the plasma.

Proteins

Carbohydrates

Fats

Non-protein nitrogenous substances

Internal secretions

Enzymes

Anti bodies

### **Inorganic substances of the plasma**

Following are the inorganic substances present in plasma.

1. Sodium
2. Calcium
3. Potassium

4. Magnesium
5. Chloride
6. Iodide
7. Iron
8. Phosphates
9. Copper

### **Blood cells**

1. Red blood cells or Erythrocytes
2. White blood cells or leukocytes and
3. Platelets or thrombocytes

### **Red blood cells**

Erythrocytes or red blood cells (RBC) are the non nucleated formed elements in the blood. The red colour of these cells is due to the presence of the colouring matter haemoglobin in these cells. The word 'erythros' means 'red'.

### **Normal value**

The red blood cell count ranges between – 4 to 5.5 millions per cubic millimeter of blood.

In adult males – 5 millions / cu mm of blood and

In adult females – 4.5 millions / cu mm of blood

### **Normal shape**

Normally the red blood cells are disc shaped and biconcave (dumb bell shaped)

## **Erythropoiesis**

Erythropoiesis is the process by which the origin, development and maturation of erythrocytes occur.

Hemopoiesis is the process, which includes origin, development and maturation of all the blood cells.

### **Site of erythropoiesis**

In postnatal life and in adults , the red blood cells are produced only in the red bone marrow.

1. Up to the age of 5 to 6 years : The red blood cells are produced in red bone marrow of all bones.
2. From 6th year to the 20th year : The red blood cells are produced by long bones and membranous bones.
3. After the age of 20 years : The red blood cells are produced from all membranous bones like vertebra, sternum, ribs, scapula, iliac bones. After 20 years of age, the shaft of the long bones becomes yellow bone marrow because of fat deposition and losses the erythropoietic function. During disorders of bone, the red blood cells are produced in spleen.

### **Stage of erythropoiesis**

1. Proerythroblast
2. Early normoblast
3. Intermediate normoblast
4. Late normoblast

5. Reticulocyte and
6. Matured erythrocyte

### **Stem Cell Series**

Uncommitted pluripotent hemopoietic stem cell



Committed pluripotent hemopoietic stem cells



Colony forming blastocytes (CFB)



Colony forming unit E (CFU – E)



Proerythoblast



Early normoblast



Intermediate normoblast



Late normoblast



Reticulocyte



Matured erythrocyte

### **Proerythroblast (Megaloblast)**

This is the first cell derived from the stem cell. The proerythroblast does not contain hemoglobin and it is also called the megaloblast. The proerythroblast multiplies several times and finally forms the cell of next stage called early normoblast.

### **Early normoblast**

In the nucleus, the nucleoli disappears. The cytoplasm is basophilic in nature. So this cell is also called basophilic erythroblast.

### **Intermediate normoblast**

The nucleus is still present. The hemoglobin starts appearing.

### **Late normoblast**

Nucleus becomes small with very much condensed chromatin network and it is known as ink spot nucleus.

In last normoblast, the nucleus disintegrates and disappears. The process by which nucleus disappears is called karyorrhexis.

### **Reticulocyte**

This is otherwise known as immature red blood cell.

### **Matured Erythrocyte**

Now the reticular network starts disappearing and the cell becomes the matured red blood cell.

The matured red blood cell is biconcave and it is smaller in size with a diameter of 7.2 microns. It contains hemoglobin and lacks nucleus.



It takes 7 days for the formation and maturation of red blood cells.

### **Factors necessary for erythropoiesis**

Various substances are necessary for the development and maturation of erythrocytes. These factors are classified into 3 categories namely,

- General factors
- Maturation factors and
- Factors necessary for hemoglobin formation

#### **General factor**

1. Erythropoietin
2. Thyroxine
3. Growth inducers like interleukin 3 and
4. Vitamin B
5. Vitamin C and
6. Vitamin D

### **Hemoglobin**

Hemoglobin is a conjugated protein. It consists of a protein combined with an iron containing pigment.

i.e., globin + heme  $\Rightarrow$  hemoglobin

### **Synthesis**

Each heme molecule combines with one globin molecule to form hemoglobin. The heme portion of hemoglobin is synthesized in mitochondria. The protein part globin is synthesized in ribosomes.

**Necessary factors**

1. First class proteins, the proteins of high biological value.
2. Metals – Iron, copper, cobalt and nickel
3. Vitamins – C, riboflavin, nicotinic acid and pyridoxine

**Destruction**

After the life span of 120 days, the red blood cell is destroyed in the reticuloendothelial system particularly in spleen and the hemoglobin is released into plasma.

**White blood cells**

Leukocyte are the colourless and nucleated, formed element of blood. It plays very important role in defence mechanism of the body.

**Granulocytes**

- Neutrophils
- Eosinophils
- Basophils

**Agranulocytes**

- Monocytes
- Lymphocytes

## **Morphology**

### **Neutrophils**

Polymorphs have fine or small granules in the cytoplasm, granules are violet in colour. The nucleus is multi lobed.

### **Eosinophils**

Eosinophils have coarse granules. The nucleus is bilobed. The granules stain bright red or orange with eosin.

### **Basophils**

Basophils also have larger granules in the cytoplasm. The granules stain purple blue with basic dyes. Nucleus is bilobed.

### **Monocytes**

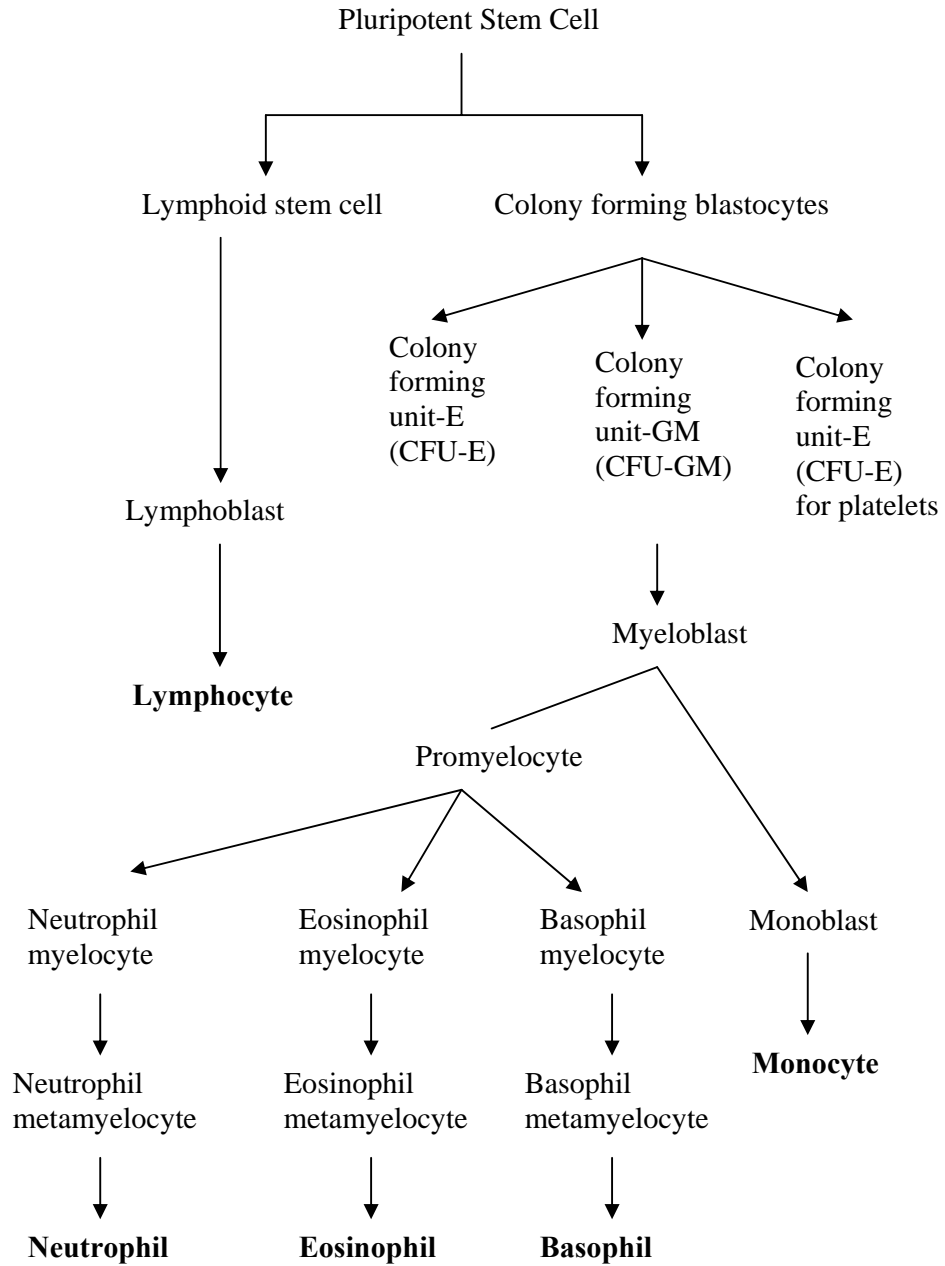
- Largest leukocytes
- Without granules
- The nucleus is round, oval, horseshoe or kidney shaped
- Nucleus is in the center of the cell

### **Lymphocytes**

- less amount of cytoplasm
- nucleus is oval or kidney shaped occupying the whole of the cytoplasm.

### **Leukopoiesis**

Leukopoiesis is the development and maturation of leukocytes.



## **Platelets**

Thrombocytes are

- Small colourless
- non nucleated
- refractive bodies

These formed elements of blood are considered to be the fragments of cytoplasms

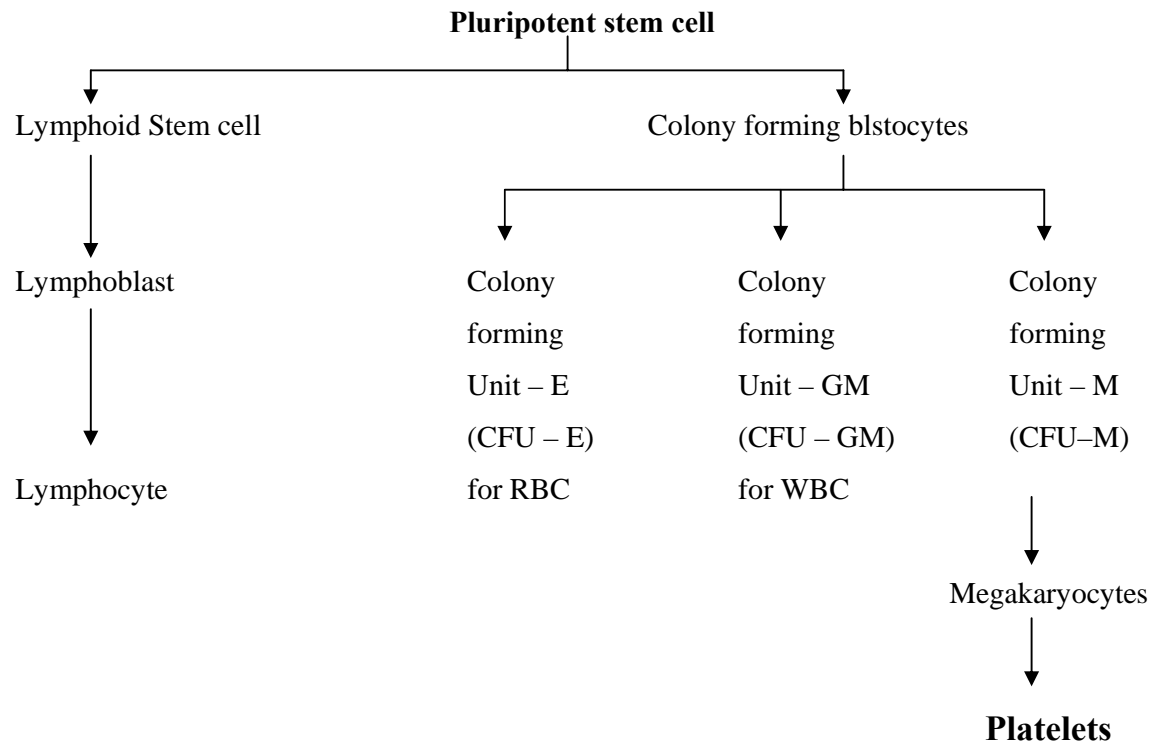
**Diameter** : 2.5 microns

**Volume** : 7.5 cubic microns

**Shape** : Spherical (or) rod shaped

### **Development of platelets**

Platelets are formed from bone marrow. The stem cell, pluripotent stem cell gives rise to the CFUM. This develops into megakaryocyte. The cytoplasm of megakaryocyte forms pseudopodium. A portion of pseudopodium is detached to form platelet, which enters the circulation



## Structure and composition

Platelets have a cell membrane, microtubules below the cell membrane and the cytoplasm

## Cell membrane

It is 6 nm thick and contains

- Lipids (Phospholipids, cholesterol and glycolipids)
- Carbohydrates (Glycocalyx)
- Proteins (Glycoproteins)

## Glycoproteins

It prevents adherence of platelets to normal endothelium but accelerate the adherence of platelets to collagen and damaged endothelium in ruptured blood vessels. It also forms receptors for ADP and thrombin.

## **Phospholipids**

Phospholipids accelerate the clotting reactions. These are the precursors of thromboxane A<sub>2</sub> which is a prostoaglandin.

## **Microtubules**

These are made up of polymerized proteins called tubulin. The tubules provide a structural support for the inactivated platelets to maintain the disc like shape.

## **Cytoplasm**

The cytoplasm of the platelets contains golgi apparatus, endoplasmic reticulum, micro tubules, micro vessels, filaments and different types of granules.

Cytoplasm also contains some chemical substances like proteins, enzymes, hormonal substances etc.

## **Proteins**

The major proteins of platelets are contractile proteins responsible for contraction of platelets

These are

- Actin
- Myosin
- Thrombosthenin

Other proteins are

- Von wille brand factor – responsible for adherence of platelets
- Fibrin stabilizing factor – a clotting factor

- Platelet derived growth factor (PDGF) – involved in repair of damaged blood vessels.

### **Enzymes**

- adenosine triphosphate
- adenosine diphosphate (ADP)
- Prostaglandins

### **Hormones**

- adrenaline
- 5HT (Serotonin)
- Histamine

### **Other substances**

- Fibrin – Stabilizing factor
- growth factor

### **Properties**

#### **1. Adhesiveness**

When platelets come in contact with any wet surface or rough surface, these are activated and stick to other surface.

Adhesiveness are

- Collagen
- Thrombin
- ADD
- Thromboxane A<sub>2</sub>



- Calcium ions and
- Vonwille-brand factor

## **2. Aggregation (grouping of platelets)**

The activated platelets group together and become sticky. The stick ness is due to

- ADD and
- Thromboxane A<sub>2</sub>

## **3. Agglutination**

It is the clumping together of platelets. It occurs due to the action of platelet agglutinins.

### **Life spam of platelets**

- 10 days it varies between 8 and 11 days
- destroyed by tissue macrophage system in spleen

### **Functions of platelets**

Normally, the platelets are inactive and execute their actions only when activated.

### **Role in blood clotting**

The platelets are responsible for formation of the intrinsic prothrombin activator. This substance is responsible for the onset of blood clotting.

**Role in clot retraction**

In the blood, the blood cells including platelets are entrapped in between the fibrin threads. The cytoplasm of platelets contains the contractile proteins namely actin, myosin and thrombosthenin. The contractive proteins are responsible for clot retraction.

**Role in prevention of blood loss (Hemostasis)**

Platelets accelerate the processes of hemostasis by three ways.

- a. Platelets secrete 5 HT, which causes the constriction of blood vessels.
- b. Due to the adhesive property, the platelets can seal the damage in blood vessels like capillaries.
- c. They play important role in plug formation

**Role in repair of ruptured blood vessel**

The platelet derived growth factor (PDGF) formed in cytoplasm of platelets is useful for the repair of the endothelium and other structures of the ruptured blood vessels.

## **Hemostasis**

When a blood vessel is cut or damaged, the injury initiates a series of reactions, which prevent further blood loss. This type of arrest of bleeding is called hemostasis.

### **Stages of hemostasis**

1. Vasoconstriction
2. Formation of platelet plug and
3. Coagulation of blood

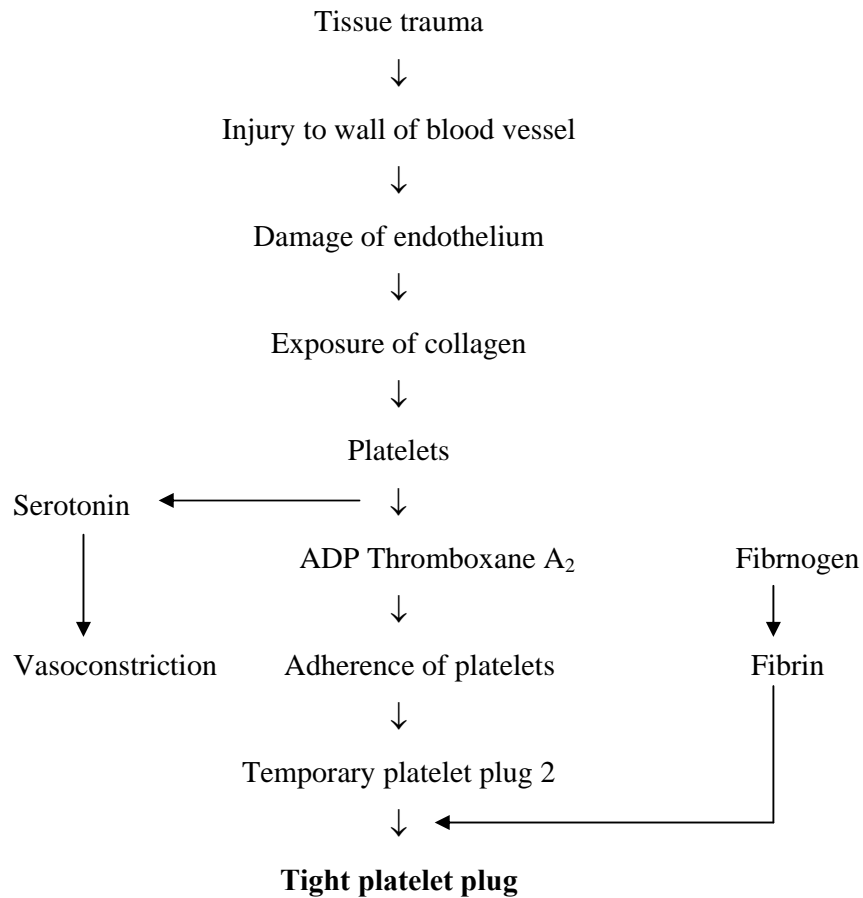
#### **1. Vasoconstriction**

Immediately after the cut, there is constriction of the blood vessel and this decreases loss of blood from the damaged vessel. Usually arterioles and small arteries constrict. The vasoconstriction is purely a local phenomenon. When the blood vessels are cut, the endothelium is damaged and the collagen is exposed. When platelets adhere to this collagen, they secrete serotonin and other vasoconstrictor substances. These substances cause constriction of the blood vessels.

#### **Formation of platelet plug**

When platelets adhere to the collagens of a ruptured blood vessel, these platelets secrete ADP and thromboxane  $A_2$ .

These two substances attract more and more platelets and activate them. So more platelets aggregate and form a temporary loose plug, which closes the vessel and prevents the blood loss.



## Blood coagulation

When blood is shed out or collected in container, it loses its fluidity and becomes a jelly like mass after few minutes. This process is called coagulation or clotting of blood. The clot is a mesh of these fibrils entangling the blood cells. These fibrils consist of fibrin. The fibrin is formed from fibrinogen.

## Basic theory

More than 50 important substances that affect blood coagulation have been found in the blood and tissues.

Substances that promote coagulation are called procoagulants

Substances that inhibit coagulation are called anticoagulants

The blood will coagulate depending on the balance between these two groups of substances.

The anticoagulants normally predominate, and the blood does not coagulate, but when a vessel is ruptured, procoagulation in the area of damage become “activated” and override the anticoagulants, and then a clot develops.

### **Factors involved in blood clotting**

Coagulation of blood occurs through a series of reactions due to the activation of a variety of substances. These substances necessary for clotting are called clotting factors.

The clotting factors are

<b>Clotting factor</b>	<b>Synonyms</b>
Factor I	Fibrinogen
Factor II	Prothrombin
Factor III	Tissue thromboplastin
Factor IV	Calcium
Factor V	Proaccelerin, labile factor, Ac-globulin (AC-G)
Factor VII	Serum prothrombin conversion accelerator
Factor VIII	Antiohemophilic factor (AHF), anti-hemophilic globulin (AHG), anti hemophilic factor A
Factor IX	Plasma thromboplastin component (PTC) Christmas factor anti hemophilic factor B.

Factor X	Stuart factor, stuart prower factor
Factor XI	Plasma thromboplastin antecedent (PIA) antihemophilic factor C.
Factor XII	Hageman factor
Factor XIII	Fibrin stabilizing factor
Prekallikrein	Fletcher factor
High molecular weight kininogen	Fitzgerald factor, HMWK
Platelets.	

### **Sequence of clotting mechanism**

Normally during circulation, the blood does not clot, because the enzymes involved in clotting are in inactive form. Slight initial activation causes clotting in which each enzyme activates another one in a sequential manner till the conversion of fibrinogen into fibrin occurs.

### **Stages**

Stage 1. Formation of prothrombin activator

Stage 2. Conversion of prothrombin into Thrombin

Stage 3. Conversion of fibrinogen into fibrin.

### **Stage 1. Formation of prothrombin activator**

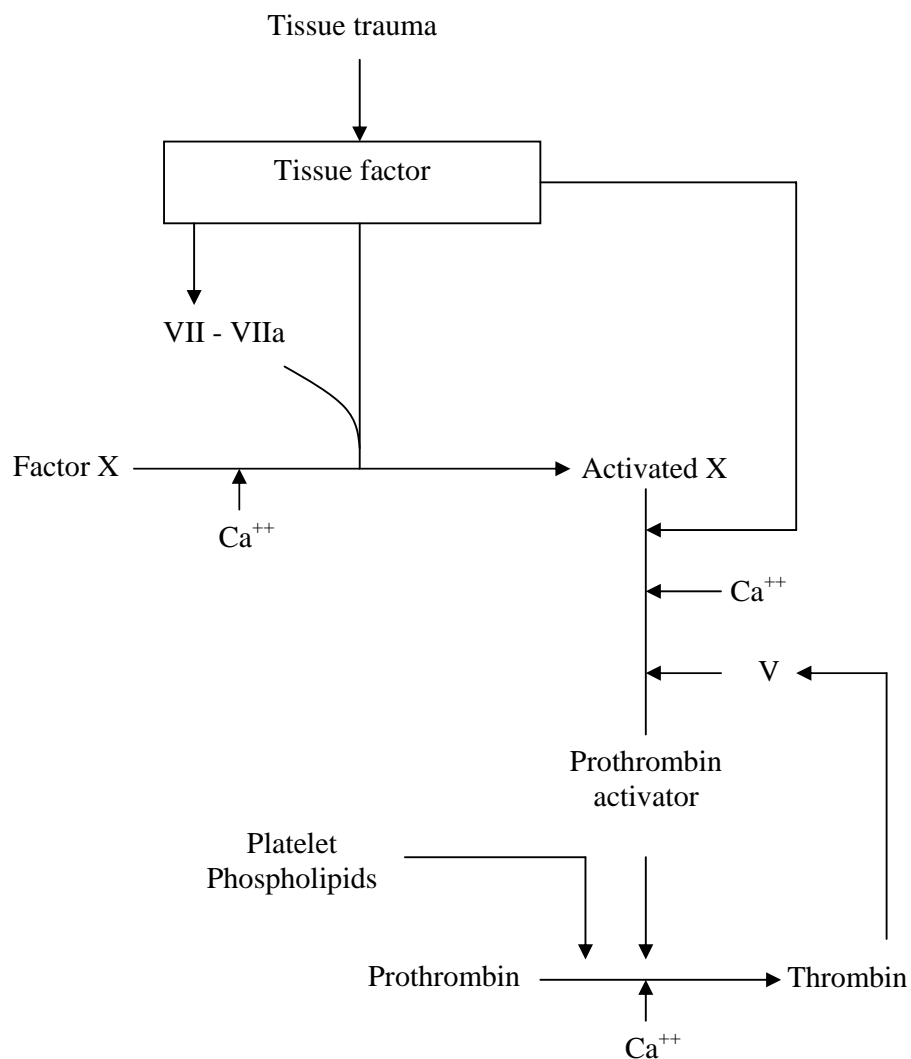
Prothrombin activator is formed in two ways,

- Extrinsic pathway
- Intrinsic pathway

## Extrinsic mechanism for initiating clotting

The extrinsic mechanism for initiating the formation of prothrombin activator begins with a traumatized vascular wall or extra vascular tissues and occurs according to the following steps.

1. Release of tissue factor
2. Activation of factor X-role of factor XII and tissue factor
3. Effect of activated factor X to form prothrombin activator role of factor V.



### **Release of tissue factor**

Traumatized tissue release a complex of several factors called tissue factor or tissue thromboplastin.

This is composed of

- Phospholipids
- Protein
- Lipo protein

### **Activation of factor X**

The lipo protein complex of tissue factor further complexes with blood coagulation factor VII and in the presence of calcium ions, acts enzymatically on factor X to form activated factor X.

### **Formation of prothrombin activator**

The activated factor X combines immediately with tissue phospholipids or with additional phospholipids released from platelets as well as with factor V to form the complex called prothrombin activator. This reaction requires the presence of calcium ions. Factor V could be activated by thrombin formed from prothrombin. This factor V now accelerates formation of prothrombin activator and the other processes of blood clot. This effect of thrombin is called positive feed back effect of thrombin.

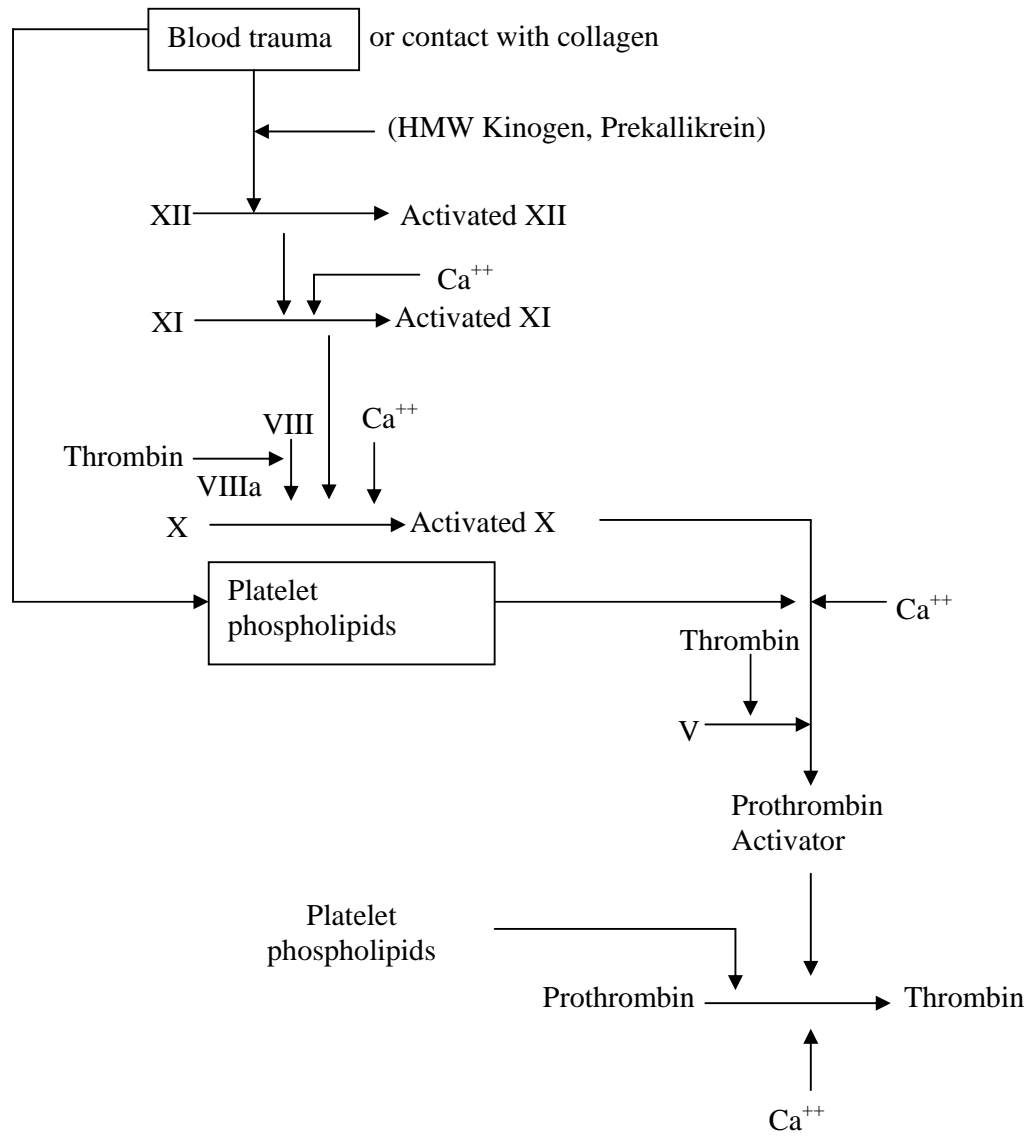


### **Intrinsic mechanism**

The intrinsic mechanism for initiating the formation of prothrombin activator, and therefore for initiating clotting begins with trauma to the blood itself or exposure of the blood to collagen in a traumatized blood vessel wall and then continues through the following series.

1. Blood trauma causes activation of factor XII and release of platelet phospholipids
2. Activation of factor XI
3. Activation of factor IX
4. Activation of factor X
5. Action of activated factor X to form prothrombin activator, role of factor V.

## Intrinsic pathway



### 1. Blood trauma causes activation of factor XII and release of platelet phospholipids

Trauma to the blood alters two important clotting factors in the blood,

- Factor XII and
- Platelet

When factor XII is disturbed, it comes into contact with collagen or with a wettable surface such as glass, that converts it into a proteolytic enzyme called “activated factor XII”.

Simultaneously, the blood trauma also damages the platelets because of adherence to either collagen or a wettable surface and this releases platelet phospholipid that contains the lipoproteins called platelet factor 3, which also play a role in subsequent clotting reactions.

## **2. Activation of factor XI**

The activated factor XII converts factor XI in to activated factor XI in the presence of kinogen and prekallikrein.

## **3. Activation of factor IX**

The activated factor XI activates factor IX in the presence of calcium.

## **4. Activation of factor X : role of factor VIII**

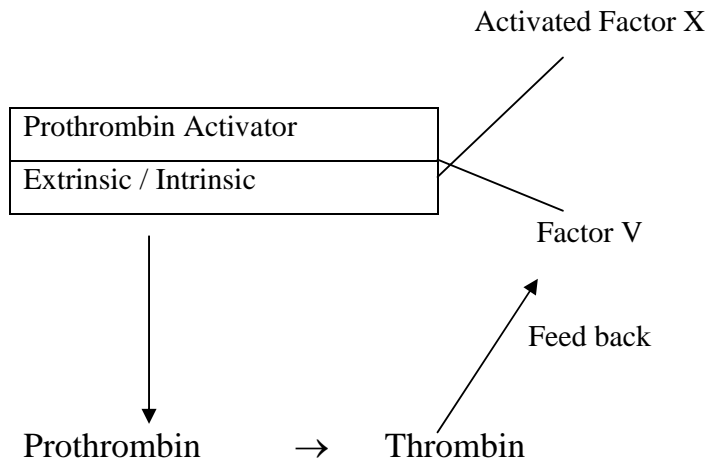
Activated factor IX activates factor X in the presence of factor VIII and calcium.

It is clear that when either factor VIII or platelets are in short supply, this step is deficient. Factor VIII is the factor that is missing in the person who has classic **haemophilia**, for which reason it is called **antihaemophilic** factor.

## **5. Formation of prothrombin activator – role of factor**

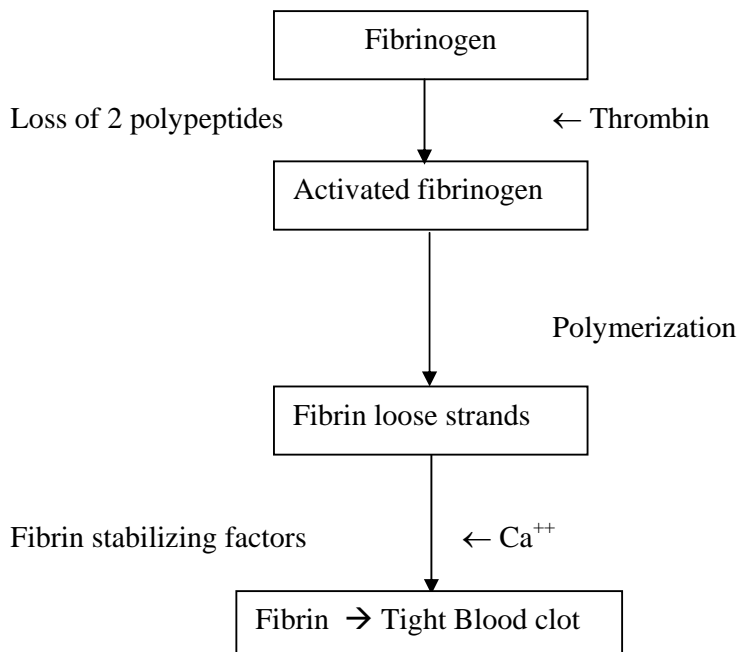
Activated factor X combines with factor V and platelet of tissue phospholipids to form the complex called prothrombin activator.

## Stage II: Conversion of prothrombin in to thrombin



Prothrombin activator converts prothrombin into thrombin in the presence of calcium. Thrombin itself accelerates this reaction by positive feed back mechanism. That is, the initial thrombin activates factor V. This in turn accelerates the formation of both extrinsic and intrinsic prothrombin activation.

## Stage 3 Conversion of Fibrinogen into Fibrin



During this process, the soluble fibrinogen is converted into in soluble fibrin by thrombin. Initially the firbinogen is converted into activated fibrinogen due to loss of 2 pairs of poly peptides from each fibrinogen molecule. The activated fibrinogen is called fibrin monomer. This p.lymeriges with other monomer molecules to form fibrin.

The first formed fibrin contains loosely arranged strands. This is modified later into a dense tight aggregate by fibrin stabilizing factor (XIII) and this reaction requires the presence of calcium ions.

### **Blood clot**

The fibrin threads run in all directions. The red blood cells, white blood cells and the platelets gets entrapped within the meshwork of fibrin. The entire mass of fibrin meshwork and the blood cells entrapped within this is called blood clot. The blood clot adheres to the opening of damaged blood vessel and prevent blood loss.

## **Genetics**

Genetics is the science that deals with the transmission of characters from parents to off spring. An elementary knowledge of the principles of genetics is essential for under standing the causation of several diseases.

### **Chromosomes**

During the cell division, the chromatin network in the nucleus becomes condensed in to a number of chromosomes.

### **Structure of chromosome**

Chromosomes are made up of two rod-shaped structures or chromatids placed more or less parallel to each other. The chromatids are united to each other at a light staining area called the centromere. Each chromatid has two arms, one on either side of the centromere.

### **Significance of chromosomes**

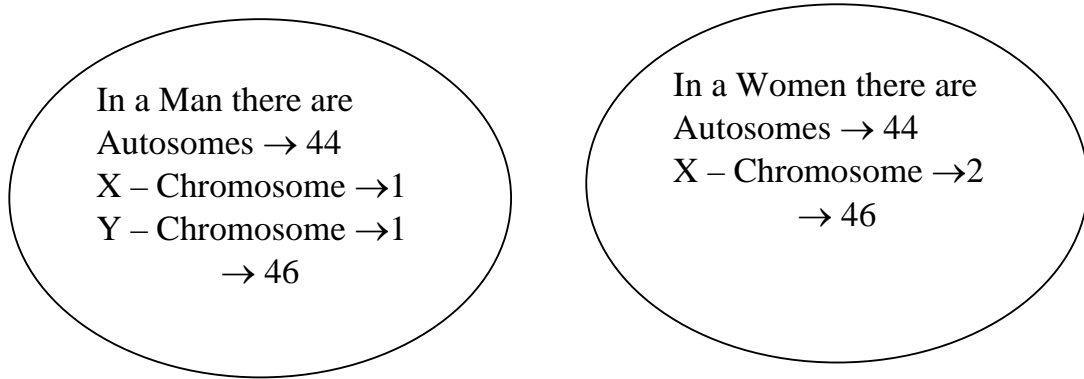
- Chromosomes are made up of predominantly of a nucleic acid called deoxyribonucleic acid (DNA)
- All information is stored in molecules of this deoxyribonucleic acid
- When the need arises, this information is used to direct the activities of the cell by synthesizing appropriate proteins.

### **Haploid and diploid chromosomes**

- The number of chromosomes in each cell is fixed for a given species.
- Man – 46 chromosomes. This is referred to as the diploid number.

- In spermatazoa and ova the number of chromosomes is only half the diploid number. ie 23. This is called the haploid number

Number of chromosomes in somatic cells of a man and of a woman.



## Genes

- Each chromosome bears on itself a very large number of structures called genes.
- These guide the performance of particular cellular function, and in turn may lead to the development of particular features of species or of an individual.
- Gene is responsible for transmission of a particular character from parents to off spring.
- Every gene occupies a definite position on the chromosome to which it belongs. This position is called the locus of the gene.

The two chromosomes of a pair carry genes that are identical in number, and in the sequence in which they are arranged. It therefore follows that the

genes of an individual are arranged in pairs, one gene being derived from the father and the other from the mother.

During gametogenesis the two chromosomes of each pair, again separate and pass to different gametes. The chromosomes that were originally derived from the father, and those that were derived from the mother, are distributed to gametes in a purely random manner. As a result the various gametes produced by an individual carry different combinations of chromosomes and therefore, of genes. When these gametes take part in fertilization, new chromosome pairs are formed.

### **Homozygous**

If the two genes are alike the individual is said to be homozygous.

### **Heterozygous**

If the two genes of the pair are dissimilar the individual is said to be heterozygous.

### **Dominant character**

The character which is capable of expression when carried by only one of a pair of homologous chromosomes.

### **Recessive character**

The character which is incapable of expression unless the responsible allele is carried by both members of a pair of homologous chromosomes.

### **Recessive inheritance**

The characteristics of a disease transmitted as a recessive character.



1. Diseased individual generally have parents who are apparently normal.
2. More than one brother or sister may be affected
3. The abnormality is more common, seen in children resulting from marriages between close relatives
4. Diseased persons who marry normal individuals usually have normal children.

A disease persons is likely to have affected children if he marries a close relative

5. If two diseased persons marry all their children are affected.

### **Mutation**

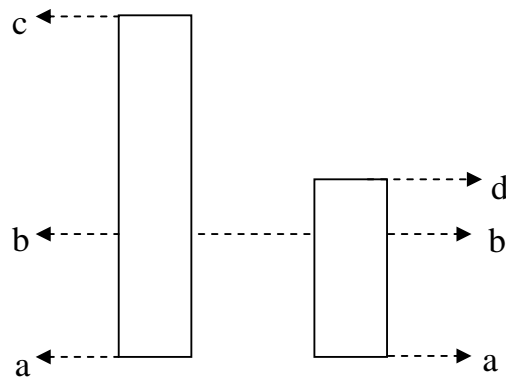
Genes sometimes undergo physico – chemical changes that alter their effect on the character controlled by them. Such a change is called mutation. The mutant gene is transmitted to future generation just like the original gene. The new gene formed by mutation occupies the same locus, upon the same chromosome, as the original gene.

### **Mutant genes**

A gene is a part of DNA molecule that carries the code for the synthesis of a single polypeptide chain. The code is dependent on the sequence of bases on this part of the DNA molecule. When a mutation occurs this sequence is disturbed. This alters the sequence of amino acids in the protein synthesized. Replacement of even one amino acid by another can change the entire nature of the protein concerned and may lead to profound effects.

## Sex-linked inheritance

Sex chromosomes carry genes responsible for the determination of sex. They also carry other genes that control characters that have nothing to do with sex. However, inheritance of characters carried by these genes may be apparent only in one sex and not in the other.



The human X and Y chromosomes are shown diagrammatically. The greater part of the X chromosome (from back) is not homologous with Y. Thus genes present on this part of the X – Chromosome are not present in Y. It follows that, in a male there will be only one set of these genes (and not pairs as in the case of other genes). As no crossing over can occur between this region of the X Chromosome and the Y chromosome, genes present here always move with the X Chromosome. They are said to be totally linked to the X Chromosome. Almost all conditions that are described as sex linked are due to genes on this part of the X Chromosome. The parts a to b of the X and Y chromosomes are homologous and genes located here are said to be partially sex linked. Finally there is the part b d of the Y chromosome which is not homologous with any part of the X chromosomes. Sex linkage in relation to

segments ab and bd is rare in man. In the account that follows, only total linkage to the X chromosome is considered.

The male has only one X chromosome. As a result, genes carried on it are unpaired and there is no question of the character controlled by the gene being dominant or recessive. If the gene is abnormal, the abnormality will always manifest itself.

The female, on the other hand, has two X chromosomes. Therefore, we may have dominance recessiveness or intermediate manifestations just as in character carried by autosomal genes.

Haemophilia is a classical example of a disease produced by a recessive gene linked to the X Chromosome. A male has only one gene at this locus. If the gene is normal the person is normal, if the gene is abnormal he suffers from hemophilia.

A female has two genes at the locus. When both of these are normal the woman neither manifests hemophilia nor transmits it. If one gene is abnormal and the other normal she still does not manifest the disease (as the abnormal gene is recessive) however, she can pass on the abnormal gene to her children. If both genes are abnormal, she will suffer from hemophilia.

A female hemophilic can be produced only if both parents suffer from the condition, or if the father is a hemophilic and the mother a carrier (heterozygote). Both possibilities are remote. This explains why the disease is very uncommon in women.

## **Haemophilia**

Haemophilia was the first haemorrhagic disorder to be accurately described when it was recognised as a hereditary bleeding disorder of males which is transmitted by healthy women. Until 1952, it was thought that the coagulation abnormality in haemophilia was always caused by the deficiency of the blood clotting factor antihæmophilic factor (AHF or F VIII) but it was then that the blood of some patients was deficient in a previously unrecognised clotting factor Christmas factor (PTC or factor IX)

### **Incidence**

One in 10,000 males born with deficiency or dysfunction of the factor VII molecule and hence the heterozygous female may also occur with about the same frequency.

### **Inheritance**

In this condition, the clotting of shed blood is greatly slowed & abolished leading to serious haemorrhage. Generally, it affects males but it is transmitted by females. The father of hæmophilic individual is usually unaffected because his X chromosome carries the normal allele for the elaboration of anti hæmophilic globulin. The Y chromosome does not carry genes concerned with clotting. The mother of a hæmophilic male is heterozygous the gene for

haemophilia is often called sex-linked lethal for it may often cause death due to bleeding.

Haemophilia A and haemophilia B are genetically unrelated but both are inherited as sex-linked recessive characters because the genes for the disorders are carried by the X-Chromosomes. Thus female carrying a gene for haemophilia on one of their two X chromosomes transmit the gene to half of their female offspring and to half of their male offspring according to the laws of Mendel. On the other hand, haemophiliacs having only one X chromosome, transmit the gene to all their female offspring, but to none of their male children.

Male, who inherit a gene for haemophilia invariably have deficiency of the corresponding coagulation factor. The concentration of the clotting factor in their plasma ranges from complete deficiency to as high as 30 percent of normal. Related males with haemophilia have a similar, and usually identical, concentration of the deficient clotting factor in their blood; it does not vary with age. It can therefore be concluded that in addition to the two types of haemophilia being genetically distinct, the plasma concentration of the clotting factor concerned is also genetically controlled, and that there are different grades of haemophilia.

In about 30 percent of cases, usually with severe haemophilia, evidence of inheritance is lacking. This is presumably due to recent gene mutation which

causes the mother to become a carrier or alternatively after several generations of carrier daughters.

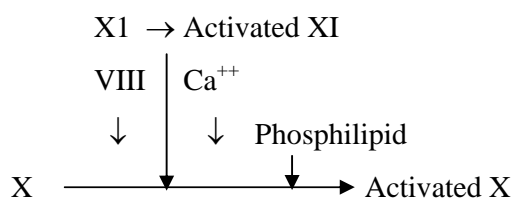
Females carrying a gene for haemophilia are called carriers. A small proportion of carriers especially of haemophilia B have a very mild tendency to bleed and a subnormal concentration of the relevant clotting factor in their blood. Rarely, the deficiency of the clotting factor is sufficiently severe to cause a moderate bleeding tendency.

Women in families with haemophilia often wish to know if they are carriers. When tests show a discrepancy between factor VIII activity and vonwillebrand factor levels, a woman can be suspected as a carrier of haemophilia A. The same for haemophilia B and in neither condition does a normal result exclude the possibility of the carrier state.

## Types of inheritance

### Haemophilia A

Haemophilia A is due to a deficiency of coagulation factor VIII which functions in the coagulation system as a cofactor with activated factor IX, phospholipid, and calcium in the activation of factor X.



## **Factor VIII**

The antihemophilic factor (AHF) or factor VIII coagulant protein, is a large, single chain protein that regulates the activation of factor X by proteases generated in the intrinsic coagulation pathway. It is synthesized in liver parenchymal cells and circulates complexed to the von Willebrand factor protein. Previous effort to purify and characterize the factor VIII molecule were limited by its low concentration and susceptibility to proteolysis. The cloning and sequencing of complementary DNA (cDNA) encoding the factor VIII molecule and the mapping of factor VIII gene on the X chromosome have provided a detailed picture of structure and have led to improved methods for carrier detection and prenatal diagnosis of haemophilia A.

### **Classification**

Based on factor VIII activity.

**Severe disease** → Patients with < 1 percent factor VIII activity will bleed frequently even without discernible trauma

**Moderate disease** → Patients with level between 1 and 5 percent will have less frequent bleeding episodes

**Mild disease** → Patients with levels over 5 percent will have infrequent bleeding that is usually secondary to trauma

Occasional patients with factor VIII levels as high as 25 percent are discovered when they bleed after major trauma or surgery. The majority of patients with haemophilia A have factor VIII levels below 5 percent.

## **The genetic basis of haemophilia A**

The vast majority of patients with haemophilia A have low or undetectable plasma levels of factor VIII protein only in crm + patients defective protein can be identified and even in these cases, analysis of the small amount of protein obtained has proved to be extremely difficult. Our present knowledge concerning the defects causing haemophilia A is therefore almost entirely based on analysis of the factor VIII gene.

As with all gene analysis, progress is related to both technological advances and to the size and complexity of the gene itself. Since the factor VIII gene was cloned and DNA probes became available, a steady stream of mutations has been reported, and are now included on a database that is updated annually. Southern blotting utilizing gene specific probes has shown an incidence of gene deletions larger than 50 bp of 2 - 5 percent of haemophilia A patients over 59 different large deletions have been reported, all, but three of which result in severe disease. Patients with a gene deletion have a five fold, greater chance of developing an inhibitor to factor VIII as a result of replacement therapy from patients with severe haemophilia without deletions.

Advances in the detection of point mutations, small insertions and small deletions are based upon extensive use of polymerase chain reaction (PCR) amplification of specific gene sequences combined with rapid mismatch detection methods in which an amplified sequence from the patient's gene can be matched against the normal sequence. Complete analysis of all 26 exons



within factor VIII gene by these methods is difficult, but it is possible to analyse at least the coding sequence of genes by reverse transcription of mRNA to cDNA followed by PCR amplification and analysis. The discovery of low levels of factor VIII mRNA in peripheral blood leucocytes has resulted in such a procedure being used to analyse the factor VIII gene from patients with haemophilia.

Over 30 percent of point mutations are at CpG dinucleotide sequences, where a C to T transition has occurred. It is now recognized that such a transition is the most common form of mutation within the human genome and probably results from methylation and spontaneous deamination of cytosine to thymidine.

In mild or moderate haemophilia A, mutations are found in almost all cases, in severe cases no mutation has been found in almost half of patients studied. In these last cases, however, there is an apparent deletion within intron 22, which leads to a failure in transcription across this intron and recently this has been shown to result from an inversion of a section of the X-Chromosome at the tip of the long arm. This inversion results in separation of the factor VIII gene into the cause of haemophilia in almost half of severe cases.

### **Haemophilia B**

Haemophilia B is an X-linked deficiency of factor IX and is clinically indistinguishable from haemophilia A. Factor IX functions at the same point in the coagulation system as factor VIII but as a serine protease, which when

activated, activates factor X - in the presence of activated factor VIII, calcium and phospholipids.

### **The genetic basis of haemophilia B**

As with most genetic disease the majority of defects causing haemophilia B have been detected by gene analysis. The factor X gene is a rather simple gene and has thus lent itself to detailed analysis using the PCR based procedures. As a result, several laboratories are reporting that in practically all cases of haemophilia B a genetic mutation can be found within the factor IX gene, or within its promotor regions. Importantly, in almost all these cases the whole of the coding and related regions of the gene have been analysed thus confirming that the defect found in the causative mutation.

In some cases mutations involve CpG dinucleotides, confirming the **hot-spot** nature of this sequence. Relatively few mutations are recorded as occurring at known functional sites, although mutations at six of the twelve Gla residues are recorded, together with three mutations at the active site, serine 365, and one at the active site, Asp 269. It is clear that all nonsense mutations cause severe disease, and that most missense mutations resulting in amino acid changes dramatically affect protein stability or release.

## **Diagnosis**

### **Carrier detection and prenatal diagnosis of haemophilia A**

Family studies in an X linked disease such as haemophilia, resulting in precise assessment of carrier status and offering the option of prenatal diagnosis, can have a major impact on the medical, psychological and social consequences of the disease. It has been estimated that for each patient an average of five to six potential carriers are present in the population. Obligate carriers (daughters of haemophiliacs or mothers of a haemophilic child with a maternal history of the disease) can be identified from the family tree. Possible carriers (daughter of carriers, female relatives of haemophiliacs on his maternal side, mothers with one haemophilic son and no family history of the disease) require laboratory investigation in order to determine their carrier status. Phenotypic assessment of carrier status in haemophilia A can be made by analysis of plasma levels of factor VIII because carriers will, on average have 50 percent of the normal female level. This method will only identify some 80 percent of obligate carriers with certainty. The most precise method is to use gene analysis to identify the causative mutation within the patients factor VIII gene and to then analyse the potential carriers for the same affect. This is now possible in some cases particularly where the chromosome inversion has occurred and with improving technology is becoming increasingly feasible. Where such techniques are not available haemophilia gene tracking can be done

by DNA polymorphisms have described with varying frequencies throughout the normal population.

Over 90 percent of female are heterozygous (informative) for at least one of these polymorphism, and therefore in haemophiliaA families the affected gene can be tracked in majority of cases. Polymorphism analysis has the advantage of being technically simple, especially when PCR based and is applicable in all families irrespective of the causative gene defect. Disadvantages include occasional non informativeness, non-availability of key-family members, limited use in sporadic haemophilia, and questions of paternity. The establishment of a national database of haemophilia patients and their individual gene defects will eventually lead to direct gene defects will eventually lead to direct identification of specific defect in potential carriers, so overcoming the disadvantages with polymorphism analysis.

### **Carrier detection and prenatal diagnosis of haemophilia B**

Seven RfLPs, have been described within or directly flanking the factor IX gene, and when all are used, over 90 percent of females are heterozygous ie informative, for at least one. Because the complete DNA sequence of the gene is known all are readily detected by PCR – based, DNA amplification procedures. The advent of procedures for the detection of factor IX gene mutation that can identify the causative mutation in practically all patients is now having an impact on genetic studies of heamephilia B and RFLP analysis is

now less frequently used. The use of the polymorphism based gene tracking is still of considerable benefit to affected families.

## **Clinical features**

### **Type and site of bleeding**

#### **Wound bleeding**

Wound bleeding is the characteristic symptom of all haemophiliacs. It is usually slow and persists for day to weeks in spite of the presence of large clots. The onset of bleeding may be immediate, but is commonly delayed for hours or even days particularly in mild, haemophilia. Recurrence of bleeding after haemostasis, has apparently occurred is particularly common.

#### **Tissue bleeding**

Spontaneous bleeding may occur into almost every tissue of the body. But is more frequently in patients with severe deficiency, infrequently in moderate deficiency and rarely in mild deficiency. Injuries causing confusion, ligamentous strains, or rupture of muscle fibres result in excessive bleeding at the site of injury in all but very mild haemophiliacs.

The extent of bleeding depends on the amount of tissue damaged, the concentration of the clotting factor, and the presence or absence of an active phase of bleeding. Bleeding in to the tissues causes content from a few millilitres to several litres. The size of haematomas and the complication which arise from them are greatly reduced by early replacement therapy.

In addition to pain and swelling, they may develop fever, anorexia, leucocytosis and anaemia of moderate to severe degree.

Retro peritoneal and mesenteric bleeding is relatively common in severe deficiency, intra-abdominal bleeding is a much more frequent cause of abdominal pain than in acute appendicitis.

### **Skin**

A tendency to bruise excessively after slight injury is noted by most haemophiliacs but spontaneous bleeding into the skin and subcutaneous tissue is common only in severe deficiency. Superficial abrasions rarely cause excessive bleeding, but lacerations and contused wounds are frequently followed by prolonged troublesome bleeding lasting many weeks, petechial bleeding is rare.

### **Mouth and nose**

Bleeding from lacerations of the tongue and frenulum of the upper lip is common in children. Bleeding from the gum is uncommon during primary dentition, but is sometimes troublesome during the shedding of these teeth. Bleeding from the sockets after tooth extraction is almost invariable and in mild haemophiliacs, is often the first and sometimes the only manifestation of the disease. In severe haemophilia, spontaneous epistaxis and bleeding into the muscular tissue of the tongue are not uncommon.

### **Synovial joints**

Prior to effective replacement therapy, patients with severe haemophilia suffered from recurring haemorrhage into joint spaces. It is less likely in

moderate haemophilia. Haemarthroses may occur spontaneously, but usually result from a minor joint strain or from a direct injury. The pain and disability of a haemarthrosis depend on the rapidity and duration of bleeding, and vary from mild to very severe, commonly the acute symptoms persist for 3-4 days. In infants, the ankle joints are most commonly affected, but in older children and adults the knees are most commonly involved, while the shoulder, wrist, hip and finger joints are affected last frequently. The synovial joints of the spinal column are affected only rarely.

The presence of blood in the joint causes synovial inflammation, and respective bleeding erodes articular cartilage and causes osteoarthritis, articular fibrosis, joint ankylosis, and eventually muscle atrophy.

## **CNS**

Intracranial haemorrhage, either extra or intra cerebral, is not uncommon, particularly in severe haemophilia. In children, it occurs most commonly after head injury, but in adults, it is usually spontaneous. Bleeding into the spinal cord and canal is rare.

## **Urogenital tract**

Haematuria is a not uncommon symptom of some patients with severe haemophilia, in whom it usually occurs without apparent cause. Bleeding usually lasts for 7-14 days, and is sometimes, accompanied by ureteric colic due to the passage of clots. It is very rare in mild haemophilia, except after trauma.

## **Gastro intestinal tract**

Haematemesis and bleeding per rectum are not uncommon, but rarely occur in the absence of other symptoms, gastro intestinal bleeding is commonly the result of ingestion of aspirin or alcohol.

## **Complications of haemorrhage**

The incidence and severity of complication is greatly lessened by early and adequate treatment.

## **Pain**

Pain is the most common and disturbing symptom of haemophiliacs, particularly those with severe disease. Local pain is due to the increasing pressure in a haematoma caused by persistent bleeding. Referred pain results from the pressure effects of haematomas on peripheral nerves, and nerve roots or trunks. Visceral type pain most commonly results from intramural haematomas of the intestinal wall, occasionally, it is due to mediastinal haemorrhage.

## **Anaemia**

Frequently develops in patients with severe and moderate haemophilia, and is due to blood loss. When blood loss is acute, the anaemia is normocytic in type or is slightly macrocytic due to the increased reticulocyte count with chronic loss, hypochromic anaemia due to iron deficiency may develop.

## **Chronic haemophilic arthritis**

Permanent joint damage usually results from repeated haemorrhages into a joint, but may follow a single haemorrhage. The limitation of movement is



usually due to fibrous adhesions within the joint, but may be due to osteophytic out growth.

### **Pressure effects and sequence of haematomas**

The most serious complications of haemophilia results from the pressure of haematomas on sensitive or vital structures. Compression of peripheral nerve is common, and results in transient or prolonged paresis, anaesthesia, or hyperaesthesia. The femoral nerve and other branches of lumbar plexus are most often affected.

A condition resembling volkmann's ischaemic contracture is not an uncommon complication of a haematoma in the muscles of the forearm.

Respiratory embarrassment is a relatively uncommon but serious complications, it may result from an obstruction of the nasopharynx, by a haematoma of the tongue or from a haematoma of the larynx, due to blood spreading down to the facial planes after tooth extraction or to local anesthetic.

Recurring subcortical or medullary haemorrhage in bones leads to the formation of pseudotumors which slowly increase in size over years.

Haemophilic bleeding occurs hours or days after injury, can involve any organ, and if untreated, may continue for day or weeks. This can result in large collections of partially clotted blood putting pressure on adjacent normal tissues and can cause necrosis of muscle-compartment syndromes. Venous congestion - pseudo phlebitis or ischemic damage to nerves.

## **DISCUSSION**

Yugimuni has classified diseases into two types. They are,

Functional disorder and

Organic invasions

The functional units of our body are the three vital forces, which are vatham, pitham and kabam. Any disturbance in the vital humour will affect the function of the organ. In chronic condition, it may lead to pathological changes in the affected organ.

Vatham is the initiator of all activities of our body. It is important in the connecting network of the body from sense organ to brain and tissue to tissue and even cell to cell.

The clinical study on all selected cases were undergone investigation by both siddha as well as modern allied parameters.

### **INTERPRETATION OF SIDDHA PARAMETERS**

#### **FAMILY HISTORY**

Most of the cases explained a positive family history of the disease

#### **AGE GROUP**

No specific age group is mentioned in this disease.

#### **SEX**

Mostly male is affected because of this disease is x- linked recessive disorder.

## **CLINICAL FEATURES**

All the patients depicted the clinical features mentioned in the poem “vaigitha vatham” in the text “yugi vaithiya chinthamani”.

### **ENVAGAI THERVUGAL- Interpretation**

#### **NAADI**

In naadi diagnosis of the all patients, the observed naadi is vatha kabam, vatha pitham and pitha vatham.

#### **SPARISAM**

Swelling and tenderness is present in affected area.

#### **NAA**

On examination some patients have paleness of tongue.

#### **NIRAM**

On observations the body colour of all the patients are normal. But the affected area is swollen and reddish black in colour.

#### **NEIKURI**

Most of the patients neikuri exhibited as the oil spreads fast and look like sievehole (°øÄ"¼î ,ñ). In some patients the oil spreads slowly and look like sievehole.

## **MUKKUTRA NILAI**

### **VATHAM**

In vaigaithavatham, affected vathams are

Viyanan – Difficulty in movements due to swelling

Udhanan – Cough present

Samanan – It's balancing function is disturbed

### **PITHAM**

In vaigaithavatham, affected pitham are

Ranjaga pitham – Paleness of the tongue

Sathaga pitham – Difficulty in doing routine works

Prasaga pitham – Colour changes in the skin at the site of injury

### **KABAM**

In vaigaithavatham, affected kabam are

Avalambagam – Balancing function is disturbed

Sandhigam – Difficulty in movements

### **UDAL THATHUKKAL**

In this, the following udal thathukkal are affected

Saaram – Dizziness, malaise

Senneer – Stagnation of blood in the site of injury

## **INTERPRETATION OF ALLIED PARAMETERS**

Suspected cases were subjected to screening test of haematology

Bleeding time

Clotting time

Prothrombin time

Partial thromboplastin time

Factor assay

Number of patients affected in bleeding time – 2

Number of patients affected in clotting time – 8

Number of patients affected in prothrombin time – Nil

Number of patients affected in partial thromboplastin time – 12

Typically all the patients have prolonged partial thromboplastin time , and also these patients were subjected to factor assay test, which confirm the diagnosis of vaigitha vatham (Haemophilia).

Number of patient's factor VIII decreased – 11

Number of patient's factor IX decreased – 1

Most of the patients have decreased amount of factor VIII and one patient has decreased amount of factor IX.

## HIGH LIGHTS OF THE DISSERTATION TOPIC

The author's dissertation topic "Vaigithavadham" comes under Vadharoga nithanam in yugi Vaidhya chinthamani.

"Å;¼Å;ö Å"¼òÐ

Âçò¼ ÅýÉçÂ;ö , ;òÐ"

If Vadha is affected, the other functions of protection and destruction are also affected.

The disease is characterized by the presence of haematoma, Pain, swelling which may involve any part of the body. The disease may also have complications like ischemic damage to nerves.

The patient complaints of swelling, which is firm in character associated with pain and restriction of day-to-day activities.

Few of them, shows the symptoms of fever, cough and neurological symptoms like numbness, probably due to compression of nerves. All of these correlates with "Vaigithavadham" explained by our great siddhar Yugeemuni.

## **CONCLUSION**

Identification of disease and its pathogenesis are pre requisite for medical practice. A detailed history taking, clinical examinations as per siddha guide lines is necessary to arrive at precise diagnosis.

The study on Vaigithavadham was carried out in this dissertation, giving importance to the characteristics of the disease like haematoma formation, pain , swelling & fever.

Diagnosis can be carried out by detailed history taking, classical clinical examination of siddha system, viz, envagaithervugal and changes in seven physical constituents and three humours.

This study on Vaigithavadham concluded that, Vaigithavadham is a bleeding disorder, “Haemophilia”, which has given relevance to modern clinical entity.

# GOVT.SIDDHA MEDICAL COLLEGE, PALAYAMKOTTAI

## NOI NADAL BRANH – V

### Annexure – 1

Name of the medical unit :

I.P.No./OP.No : Nationality :

Name : Religion :

Age : Date of Admission :

Sex : Date of Discharge :

Occupation : Diagnosis :

Income : Results :

Address : Medical Officer :

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Complaints & Duration :

H/O Present illness :

H/O Previous illness :

Personal History :

Family History :

### Clinical Examination – Siddha aspect



## **General Examination**

Yakkai :

Gunam :

Irukkai nilai :

Padukkai nilai :

Suvasa enn :

Kuruthi azhutham :

## **Special Examination**

### **Pori / Pulan**

Mei - Sensation :

Vaai - Taste :

Kan - Sight :

Mooku - Smell :

Sevi - Hearing :

### **Kanmendriyam / Vidayam**

Vaai - Vasanam :

Kai - Dhanam :

Kaal - Kamanam :

Eruvai - Visarkam :

Karuvai - Anantham :

### **Paruvakalam**

Kaarkalam :  
Koothirkalam :  
Munpanikalam :  
Pinpanikalam :  
Elavernirkalam :  
Mudhuvenirkalam :

### **Utkayam / Athakayam**

Puyam - Forearm :  
Sayam - Arm :  
Kaal - Leg :  
Paatham - Feet :

### **Uyir thathukkal**

#### **1)Vatham**

Pranan :  
Abanan :  
Viyanan :  
Uthanan :  
Samanan :  
Nagan :  
Koorman :  
Kiruharan :  
Devathathan:

Dhananjayan :

## 2) Pitham

Anilam :

Ranjagam :

Pirasagam :

Aalosagam :

Sathagam :

## 3) Kabham

Avalambagam :

Kilethagam :

Pothagam :

Tharpagam :

Santhigam :

## Ezhu Udal Thathukkal

Saaram :

Senneer :

Oon :

Kozhuppu :

Enbu :

Majjai :

Sukkilam / Suronitham :

**EN VAGAI THERVUGAL**

## **MEI KURI (SPARISM)**

### **Examination of the Skin**

#### **Inspection**

Colour of the Skin

Eruptions

Haemorrhages

Ulcers, excoriations, fissures etc.

Boils, carbuncles, scars, trophic changes etc.

#### **Eruption**

##### ***Types of rashes***

Maccular

Roseolar

Erythematous

Papular

Pustular

Lenticular

Nodular

Vesicular

Bullous

Wheals

Burrows

Blackheads

Plaques

Scales

### ***Ulcers***

Duration

Mode of onset

Associated pain

Size and pain

Nature of the floor

Character of the edge

Discharge

Tenderness

Surrounding skin

Lymphnodes

### ***Pruritis***

Infestation

Skin diseases

Metabolic & endocrine

Hepatic disorders

Renal diseases

Blood diseases

### **Examination of the hair**

Falling of the hair

Patchy loss of hair

Loss of hair in temporal region

Characteristic features of the hair

## **Sweat**

Physiological / Pathological

## **Lymphglands**

Site

Shape

Size

Consistency

Mobility

Tenderness

## **Examination of the nails**

### **Examination of the Head, neck, Face**

#### ***Skull***

Size

Shape

#### ***Face***

Eyebrows

Eye lids & Eye lashes

Nose

Lips

Ears

### *Neck*

## **Examination of the Chest**

Shape and Size

Movements

Rate of respiration

Breath Sounds : Normal / Abnormal

Heart Rate & Sounds

Examination of the Breast

Examination of the Abdomen

Shape

Size

Examination of the Genital Organs

Examination of the Extremities

***Upper & Lower Limb*** : General Examinations  
Special Examinations  
Tests for Tone, Power & reflex

## **NIRAM**

Colour of the skin, Hair, Nail, Teeth, Tongue, Gums

Sputum – Normal / Abnormal

## **MOZHI**

### ***Larynx***

Congenital

Acquired

Traumatic

### ***Tongue***

Congenital Abnormalities

***Ear*** : Deafness

***Palate*** : Cleft palate

## **VIZHI**

### **Examination of Eye**

Visual acuity

Visual field

Colour sense

### **Pupil**



Size

Equality

Regularity

Reaction of light accommodation

## **NAA**

Colour

Size

Shape

## **IRU MALAM**

### **Malam**

#### ***I. Macroscopic Examination***

Amount

Colour

Odour

Consistency

Abnormal Constituents

#### ***II. Microscopic Examination***

#### ***III. Chemical Examination***

## **Siruneer**

Quantity

Colour & Transparency

Specific Gravity

Deposit

## **NAADI**

The state of vatha, pitha and kabha naadi.

## **Examination of Pulse & its Indication**

Rate

Rhythm

Volume

Force &

Character

## **Noi kanippu**

## **MODERN ASPECTS**

## ANNEXURE – II

### General Examination

Consciousness	:	General Appearance	:
State	:	Nourishment	:
Weight	:	Facies	:
Height	:	Jaundice	:
Skin Changes	:	Engorged venis	:
Anaemia	:	Clubbing	:
Cyanosis	:	JVP	:
Pedal Odema	:	Koilonychia	:
Abdominal distension	:	Brittle Nail	:
Congential anomaly	:		
Lymphadenopathy	:		

Pluse	Rate	Rhythm	Volume	Character
			(Rt)	(Lt)
Blood Presure : mm/Hg	Upper limb	-----	-----	
	Lower limb	-----	-----	

Respiratory Rate:

### Systematic Examination

Cardiovascular System :

Respiratory System :

Gastro intestinal System :

Central nervous System :

### **Laboratory Investigations**

#### ***Blood***

TC	:	MCV	:
DC: P, L, E, B, M	:	MCH	:
Hb%	:	MCHC	:
ESR	:	Serum Protein	:
1/2hr	:	Serum Cholesterol	:
1hr	:	Blood Urea	:
RBC Count	:	Serum Iron	:
Platelet Count	:	Serum Ferritin	:
Reticulocyte Count:	:	Serum TIBC	:
PCV	:	Peripheral Blood Smear	:

#### ***Motion***

Ova :

Cyst :

Occult blood :

#### ***Urine***

Albumin :

Sugar :

Deposits :

Bile Salt :

Bile Pigment :

### **Special Investigation**

Barium meal and endoscopy :

Bone marrow examination :

Skiagram :

Sputum for AFB :

Radiological investigation :

Ophthalmoscopic examination :

E.C.G.

*Etc.* :

*Case Summary* :

*Fate of the Disease* :

*Line of treatment* :

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